THESIS

"ON CHOREA MINOR"

Presented for the Degree of M.D.

by

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"Now this affection arises from some humour falling on the nerves, and such irritation causes spasms."

Sydenham.

C H O R E A.

INTRODUCTION:

This thesis is concerned entirely with Chorea as it occurs in children. Chorea Minor as it is seen in pregnancy will be shortly referred to, but only in so far as it helps to the understanding of the pathology of the disease.

The name "Chorea" is derived from the Greek "Choreia" meaning "Dancing", though perhaps not altogether appropriate, custom has so sanctioned its use, that there is little likelihood of its ever being changed. The origin of the term is interesting, for the name was first applied to that form of hysterical religious excitement occurring during the 14th to 16th Centuries in Germany and elsewhere, which was characterised by epidemics of dancing mania. The following quaint rhyme is quoted by Hecker:-

"At Strasburg, many hundreds began
To dance and leap, both wife and man,
In open market, lane and street,
By day and night; many did not eat
Until to an end their madness came;
St. Vitus' dance they did it name.

Sufferers/

Sufferers from this condition sought relief at various shrines, and of these, that of St. Vitus, patron saint of dancers, at Zabern was the most famous, and thus the disease was often called St. Vitus's Dance, a name still in common use. By a similar process, the disease also got the name of Chorea of St. John and of St. Anthony; and in many parts of England, "St. Anthony's Dance" is the term commonly applied to Chorea in children.

Sydenham * in a brief, but vivid account of Chorea in children, transferred the name of St. Vitus's Dance from its original application to that disorder in children, which is now commonly known by his name, or by the term "Chorea Minor", and though there is no relationship between the two, except in their being in both an abnormal action of the motor system, the name has been retained.

The name "Chorea" has also been applied to many more or less allied diseases, e.g., to the various forms of motor tic, to disorders of movement preceding or succeeding hemiplegia - the so-called pre- and post-hemiplegic choreas, and to many others.

To/

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* The Works of Sydenham. Syd. Soc. translat. by Latham. Schedula Monitoria, para. 19.

To distinguish the Chorea of children from the other varieties, it is now commonly known as "Sydenham's Chorea", or "Chorea Minor".

DEFINITION:

Chorea Minor may be defined as a disease occurring mainly in children (and more especially in children in the 2nd and 3rd hemi-decades), characterised by involuntary muscular movements and by more or less impairment of the mental powers, and having a marked association with Rheumatism and Endocarditis.

ETIOLOGY:

Race: Chorea is a disease of wide distribution amongst the white races. It is rare in the negro, and Osler,* in his enquiries, could find no undoubted case as occurring in a full-blooded Indian. It is said to be unknown in the Chinese.

Heredity: A "neurotic" heredity is a very strongly predisposing cause and is found in about one sixth of the cases. Thus it is not uncommon to find that one of the parents has been insane, or epileptic, or a somnambulist, or has had chorea. The/

* "On Chorea and Choreiform Affections," by William Osler, p. 7, 1894.

The disease, besides being a hereditary one, may be in some cases a family one as well.

Risien Russell* mentions a case where the patient's father was a somnambulist, a brother epileptic and another brother a somnambulist. Osler** mentions one where both the mother and grandmother had had Chorea. Gowers*** has seen a case where a man was epileptic, his sister insane, and her two children choreic.

A heredity to Rheumatism is also frequent, and occasionally one finds a heredity both to this and to the neuroses. Clouston*=* in his "Neuroses of Development", states that he believes Chorea always to have a neurotic heredity even in rheumatic cases, and goes on to say that he constantly meets with cases in the children of his insane patients, of dipsomaniacs, and of epileptics.

Sex: The proportion between attacks in boys and in girls is given differently by different authors. Fagge =*= adding up the statistics of various/

* System of Medicine edited by Allbutt. Article, Chorea by J. S. Risien Russell, p. 830.
** Osler, loc. cit. p. 10.
*** Textbook of Diseases of the Nervous System, Gowers Vol. II., p. 591.
= Neuroses of Development (Morison Lectures, 1890), T. S. Clouston.
=*= Textbook of Medicine, Fagge and Pye Smith, Edit., 4th Vol. I., Chorea.

various authors, found that out of 1610 patients, 471 were males to 1139 females, a proportion of two males to five females. In 65 cases of Chorea treated at the Manchester Children's Hospital, Pendlebury, during the year 1902, there were 15 males to 50 females, a proportion of males rather smaller than that given above. The proportion of males attacked is greatest in early childhood and decreases rapidly after puberty.

Age: The great majority of cases occur between the ages of 5 and 20. The British Medical Association Collective Investigation Report gives nine-tenths of the cases between 5 and 20, and four-fifths between 10 and 15. If one only takes into consideration first attacks, the largest number is between 5 and 10, but including second and later attacks, the greater number is between 10 and 15. In my own cases (which belong to the latter group) there were twenty-two between 5 and 10, forty-three between 10 and 15.

Osler* arranging his cases in hemi-decades, gives the following figures:-

1st	hemi-decade	.....	33	cases,
2nd	" "	.....	228	"
3rd	" "	.....	212	"
4th	" "	.....	62	"

Cases/

* Osler, loc. cit. p. 6.

Cases in adults, except in relation to pregnancy and in old people, are rare, and in many recorded instances, the disease has not been true Chorea Minor. Trousseau, however, is quoted by Fagge as having seen a case in a woman of eighty-three.

Station in Life; The disease is very much more frequent in the poorer classes, a fact which is no doubt due to the greater strain thrown on the child's developing nervous system by bad hygienic surroundings and by less nourishing food.

Season and Climate: Very elaborate investigations into this point have been made, more especially in America by Weir Mitchell* and by Lewis, the result, however, has been indefinite. The conclusions of Lewis are (as quoted by Osler) as follows:- "No one element of 'weather' explains fully the fluctuations of these tracings for Chorea, although in the barometer and storm statistics, the relationship appears to be closer than to any other etiological factor or factors that have as yet been advanced."

I have gone over the statistics of cases of Chorea and of Rheumatism occurring in the out-patient/

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* Lectures on Diseases of the Nervous System, S. Weir Mitchell, 2nd Edit., 1885.

patient department of the Manchester Children's Hospital during 1901 and 1902 with the following result:-

CASES OF CHOREA.

In 1901 and 1902

Jan.	Feb.	Mar.	Apr.	May	June	July	Aug.	Sept.	Oct.	Nov.	Dec.	Total
40	33	36	54	29	47	56	32	65	67	45	50	554

CASES OF RHEUMATISM.

In 1901 and 1902.

Jan.	Feb.	Mar.	Apr.	May	June	July	Aug.	Sept.	Oct.	Nov.	Dec.	Total
9	5	5	16	10	8	9	7	9	8	11	7	104

It will be seen that there is a considerable rise in the number of Chorea cases during April - July, but that the maximum is attained in September and October. Comparing with this the cases of Rheumatism, we do not find any marked agreement except that here also there is a rise in April, but there is no maximum in late autumn to meet that of the cases of Chorea.

Locality: Chorea is undoubtedly a commoner disease in towns than in the country, this is in all probability due to the less healthful surroundings and to the absence of that open-air life which is so strong/

strong an antidote to any neurosis. The greater frequency of Rheumatism in towns is also an important factor.

Temperament: It is agreed by all that Chorea has a special tendency to affect bright, intelligent children with what it is customary to call "high-strung" nervous systems, or as Dr Church* aptly terms it, "the neurotic make-up". Obviously, a child of this temperament is more likely to be stimulated by ambition, or fear of punishment as the case may be, to overwork at school, and that as we shall see is by no means an unusual etiological factor in the causation of Chorea. Possibly, too, in this type of child, the motor centres as well as the intellectual and emotional, are more easily induced to energise than in his more slow-witted comrades.

Fright, Nervous Shock and Strain: The statistics of different writers vary very widely on this point. Certainly fright, as Gowers remarks, is the only common etiological factor given by the relatives, but then it is a popular belief that Chorea is caused by a fright, so the value of the/

* Nervous and Mental Diseases, Church and Peterson, 4th Edit. 1903, p. 531.

the information is small. In preparing statistics denoting the influence of fright, we must have a definite idea as to which cases can be legitimately included under this cause. As in the case recorded by Von Ziemssen*, where a boy became immediately choreic when frightened by a pistol being fired close to him, we cannot deny that fright had a definite influence, but its importance is surely exaggerated when one is asked to include cases where some weeks or even months before the child has sustained a fright. Children are easily frightened, and in many cases the alleged fright was so trivial a matter that it may be neglected.

Personally, I am not inclined to lay much stress on it. In only a very small proportion of cases admitted during 1902 to Pendlebury Hospital, was fright given as a cause, and in still fewer was it an immediately preceding cause.

In a few cases nervous shock appears to be of some importance, as for example, where Chorea has developed in a child who has recently lost his parents or other relatives.

With regard to nervous strain, there is more uniformity of opinion. Sturges** was probably one of the first to insist upon it. Out of 177 consecutive/

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* Cyclopaedia of the Practice of Medicine, Von Ziemssen, American Edition, 1877, p. 432.

** On Chorea or St. Vitus's Dance, by O. Sturges, 2nd Edition, 1893, p. 181.

secutive cases, he found that in 86 there was a history of fright, nervous shock or strain as from overwork at school and he is naturally disposed to look on these as of great importance. He gives several illustrative cases where Chorea occurred in children working hard for school examinations, and we can not doubt that any circumstances leading to exhaustion of nervous centres may have an influence in producing the disease, though possibly only in an indirect manner.

The life of the modern child at school is artificial and trying in the extreme, and there is little wonder that many children, more especially girls, at the trying period of puberty, should break down and suffer from various nervous ailments.

When one considers that many children are at school for from six to seven hours daily with very short intervals for play, that during that time they are working at high pressure and in not always perfectly lighted or ventilated class rooms, that the children are often ill fed and ill clad, and that home lessons occupy other one or two hours on their return, one does not wonder that such a system of education is productive of many evils. One does not wonder that the report of the recent Royal Commission on Physical Training in/

in Scotland has such a terrible tale to tell; one wonders rather how, under the present system, a child can find any time or energy to look after its physique in the only way it knows how, namely:- in healthy play and by means of the healthy appetite which that play develops. Surely a child's work should in all things be "short of weariness", and surely we could produce better citizens if we paid a little more attention to the needs of the body and a little less to the needs of the mind, as defined by an unduly exacting "Code".

Imitation: It may be said with confidence that true Chorea is rarely, if ever, the result of imitation, though the reverse is a common belief amongst the laity. Epidemics of Chorea supposed to be due to this cause are occasionally reported, but on careful investigation are found to be cases of Hysteria taking on some of the outward signs of Chorea. There is, however, a fairly close relationship between some cases of Chorea and some of Hysteria. Sturges * especially has endeavoured to demonstrate that the two diseases are both merely "functional" disorders of the brain, their special characteristics being due to their occurring at different epochs in the patient's life.

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* Sturges, loc. cit.

Traumatism: Is probably an occasional cause, or at least, a subsidiary one. In one case, which I have seen, choreic movements developed a few days after the child had undergone an operation for post-nasal adenoids, but considering the great number of children who undergo this operation, it may have been merely a coincidence.

Reflex Imitation: This was formerly considered to be of great importance, but this view is little held nowadays. Reflex genital irritation, reflex irritation from the presence of worms in the intestine, reflex irritation from the eyes in cases of especially hypermetropia and astigmatism, have all been often cited, but their actual influence in the production of the disease is unconvincing.

Acute Specific Diseases: Occasionally Chorea occurs shortly after an attack of one of these, but with the exception of Acute Rheumatism, their connection is probably as a rule accidental. I have seen only one case where the Chorea developed during convalescence from an acute specific disease other than Rheumatism, in this instance Scarlet Fever.

Other Diseases:/

Other Diseases: e.g., Anaemia. This is very frequently present, but is in most instances a result and not a cause.

RHEUMATISM:

The close association between Acute Rheumatism and Chorea in children has been long recognised, and its significance is still under dispute.

Statistics vary to some extent, due very largely to different ideas amongst different observers as to what constitutes Rheumatism in a child. The writings of Cheadle* have been of great service in drawing attention to the anomalous manifestations of Rheumatism in children. One can not fail to be struck after some experience of work amongst children by the rarity with which Rheumatism manifests itself amongst them in the manner which is so usual in the adult, how comparatively rarely one sees a child with polyarthritis, acid sweats, and that complete disablement which is so characteristic of Rheumatism in the adult.

On the contrary, Rheumatism in a child is often only manifested by a slight arthritis, by tonsillitis, by the occurrence of Erythema Nodosum, by endo- or peri-carditis, by "growing pains", or by subcutaneous fibrous nodules.

These/

* The Rheumatic State in Childhood. (Harvian Lectures, 1888), W. B. Cheadle.

These various terms in "the Rheumatic series" as Cheadle has termed it, may occur in any combination and in any order. Thus, a child may at one time suffer from tonsillitis and slight arthritis, at a later date from peri- or endocarditis, or the sequence may be reversed or altered in every possible way. Rheumatism in the child is not the acute process that it is in the adult, as Cheadle says "the history of a Rheumatism may be the history of Childhood".* Now, in this Rheumatic series, we find that Chorea has a place; in a large proportion of those cases of children saturated, as it were, with Rheumatism, we find Chorea occurring at one time or other, sometimes alone and at other times associated with arthritis, tonsillitis, endocarditis, or any other member of the series. As an instance of this and of the subacute progress of Rheumatism in early life, I can not do better than cite the following striking case recorded by Cheadle (loc. cit.):-

W.S., aet. 4½:

1887:-

1. Chorea, probably with Endocarditis.

Oct. 1887:-

2. After 11 months, arthritis (first attack)

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* Cheadle, loc. cit., p. 10.

Nov. 1887:-

3. Chorea, second attack; Subcutaneous nodules; Endocarditis; Erythema Marginatum.

Dec. 1887:-

4. Emotional attacks; Chorea (continued) Fresh nodules.

Jan. 1888:-

5. Erythema Marginatum; Fresh eruption of Nodules; Arthritis (second attack.)

Feb. 1888:-

6. Fresh Eruption of Erythema; Fresh crop of nodules; Tonsillitis;

March 1888:-

Death.

It will be seen that in this case the Chorea preceded the other Rheumatic manifestations; the more usual sequence is that it should succeed them or at least, some of them. An instructive case where tonsillitis, arthritis, chorea, endocarditis and iritis all occurred simultaneously is recorded in the British Medical Journal for March 7, 1903.

Turning next to actual statistics, we find the following results:-

A history of preceding Rheumatism could be found in 97 cases out of 437 cases of Chorea analysed in/

in the British Medical Collect. Investigation Report, the percentage therefore is 22%. Osler, after most careful enquiries, found it in 25 out of 144 cases, a percentage of 17.

Sturges gives 11 undoubted, 20 probable, and 17 doubtful cases out of a total of 172. Taking the undoubted and probable cases together, we have here 18%.

In my own series of 65 cases, there is a percentage of 21 with a clear history of preceding Rheumatism, and 18% with doubtful.

We may therefore take 20% as an average. A considerable number of cases, however, develop Rheumatism during an attack; the British Medical Collect. Investigation Report gives 32% as that percentage in which one gets either a history of preceding Rheumatism or Rheumatism during the attack.

In my own cases, four developed Rheumatism whilst under treatment, giving a percentage of 6, which, added to the 21%, makes 27% with Rheumatism before or during.

Batten* followed out a number of cases and found that in two years another 11.3% had developed Rheumatism, who had not previously suffered from it, and in six years the percentage amounted to 20%, and/

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* "Lancet", Nov. 5, 1898, F.E.Batten.

and his results are admittedly an under estimate. His figures are as follows:-

A history of Rheumatism present on admission 32.2%
" " " " " two yrs.later 43.6%
" " " " in other three years 53%.

Now, these figures are very striking, and when one considers that within a few years after an attack of Chorea one out of every two cases has suffered from Rheumatism either before, during, or since the attack, it is impossible to deny that the two diseases have some distinct relationship.

Chorea occurs, as a rule, subsequent to an attack of Rheumatism, but according to the British Medical Investigation on Rheumatism, it precedes it in about 2%.

Sturges found that a history of Rheumatism was commoner in young children than in old, whereas Gowers came to an exactly contrary conclusion.

In my own cases, the figures were 21.3% in children under 10, and 21.4% in children between 10 and 14, a result practically identical.

A Rheumatic family history is found in a large number of cases; Cheadle found in 86 cases that 62 (or 75%) gave satisfactory evidence of acute Rheumatism/

atism in themselves or in their immediate relations. Turning to the statistics of the age and sex incidence of Rheumatic Fever proper, it is seen that the disease is comparatively speaking, not common in children. Osler* gives only 4.3% of cases as occurring under fifteen years of age. With regard to sex, males are attacked in the proportion of 3 to 2, but up to the age of 20, females predominate, and between 10 and 15 girls are more prone to the disease. These statistics are based on the results of the British Medical Association Collective Investigation on Rheumatism. Cheadle** has still further analysed those results and gives the following figures:-

Between the ages of 11 and 15, Acute Articular Rheumatism was found in 47 girls to 25 boys, a proportion of almost two to one.

Rheumatic Heart Disease in 28 girls to 16 boys, almost 2 to 1.

Chorea associated with Rheumatism (antecedent or concurrent) in 116 girls to 32 boys, or rather more than 3 to 1.

Organic Heart Disease with Chorea in 43 girls to 13 boys, or more than 3 to 1.

Chorea generally in 131 girls to 47 boys, or almost 3 to 1.

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* Principles and Practice of Medicine, Osler, 3rd Edit., p. 167.

** Cheadle, loc. cit., p. 58, foot note.

Taking also quinquennial periods from one to twenty, he got the following results:-

From 1 to 5 years of age, Rheumatism occurred five times more frequently in boys than in girls.

From 6 to 10 years, it occurred in 15 boys to 14 girls, that is nearly equal.

From 11 to 15, it occurred in 47 girls to 25 boys, thus the proportion is suddenly reversed, for now Rheumatism is found nearly twice as often in girls as in boys.

From 16 to 20, it was found in 76 boys to 67 girls.

From these figures it is seen that Rheumatic Fever is mainly a disease of the male sex, with the exception of the one period, between the ages of 11 and 15, and as we have already mentioned, that is the period in which Chorea is most commonly found and Chorea is especially a disease of the female sex.

Recent work on the bacteriology of Acute Rheumatism brings out still more markedly its relation to Chorea, but this we shall consider later under the section of Pathology, as we shall also the significance of the relationship.

Two more factors in the etiology remain to be considered/

considered, namely:- heart disease and pregnancy.

1. Heart Disease:

Though heart disease is more usually a sequel than a precedent of Chorea, there are cases which come under one's notice, where, from the history and from the signs of cardiac hypertrophy, it is evident that the heart affection preceded the Chorea. As one would naturally expect, it is in cases with a Rheumatic history that is especially the case, and no doubt in a great majority of the remainder, the condition is due to unrecognised Rheumatism. Gowers, in forty cases of Chorea with organic heart disease, found, on first examination, that it was probable that in eighteen of those the heart affection antedated the Chorea.

2. Pregnancy:

I intend only to say a few words on this subject. The association between Chorea and Pregnancy is a very interesting one and accounts for a large proportion of cases in adults. The disease differs in no material respect from that in children. The attack certainly is often severer and of a longer duration, and Chorea Insaniens is more frequent, but/

but these are merely differences of degree and not of kind. The mortality is greater. There is the same association with Rheumatism. Buist* found a rheumatic history in 45 out of 226 cases analysed by him. A previous attack of Chorea in childhood is also of importance and was found by Buist in 66 out of his 226 cases.

Chorea Gravidarum is commonest in first pregnancies and appears to be rather more frequent in those that are illegitimate. The choreic movements generally begin about the 3rd or 4th months of pregnancy and may continue more or less all through the pregnancy and do not usually stop abruptly with delivery, but gradually die away. Spontaneous abortion was formerly thought common, but this has been shown by Wall and Andrews** to be incorrect.

The disease is usually very resistant to treatment.

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* Reports Edin. Obstet. Soc., 1894-95.

** Journal of Obstetrics & Gynaecology of the British Empire, Vol. III., p. 545.

SYMPTOMS:

It is quite unnecessary in a paper of this sort to go into full details of the symptoms of so common a disease as Chorea. It will suffice if the more striking and interesting facts be commented upon.

Chorea in its early stages is often unrecognised by the parents of the child; it may be noticed that the child is a little more irritable and wayward than usual, that she is disinclined for lessons, that she has very slight irregular movements of the face and hands, that she "fidgets", that she is apt to let things fall and is apt to upset cups, etc., at meals. This prodromal stage lasts a variable time and in acute cases may be almost absent.

A little later the disease becomes more marked and the movements can not fail to attract attention, and at last the parents recognise with alarm that the child is suffering from "St. Vitus' Dance". The disease is usually looked on with great dread by the patients' friends, more especially as there is a fairly strong opinion amongst the laity that the disease may go on to insanity. Occasionally, however, one meets a case, more usually in second or subsequent attacks, where the child is still at work or at school and where the disease is made light/

light of. I have seen a case in a miserable, emaciated girl of fifteen where the patient was still at work in a laundry, though in the fourth week of a severe attack of Chorea accompanied by Endocarditis.

In some cases an early symptom is physical prostration. Féré* describes a case of relapse where four days before any chorea was present and before the patient had been able to recognise any increased irritability, there was marked fatigue with a lessened energy of the movements; the lowered dynamographic curve showed the step-like form seen in the fatigue of hysteria and the reaction time was prolonged.

Considering first and briefly the disorders of the motor system, we find that there are:-

1. Involuntary Muscular movements.
2. Muscular weakness or even paralysis.
3. Interference with co-ordination.

1. The Involuntary Muscular Movements:

Are the most obvious feature of the disease in most cases. The movements, as a rule, are most marked in the hands, face, and tongue, and are clonic/

* "Twentieth Century Practice of Medicine", Vol. X. 1897, Chorea by Charles Féré.

clonic in type. As a rule bilateral, it is common to have them commencing on one side, and they may remain throughout the whole course of the disease so limited. Hughlings Jackson pointed out long ago that hemichorea is, however, only true as regards the limbs and not of the face, a most significant fact in determining the site of the pathological process. A true hemichorea, rigidly confined to the one side throughout the whole duration of the disease is in some cases on a different pathological footing to the ordinary variety.

Sturges made many enquiries as to the exact site where the choreic movements were first noticed, and found that the hands were the most usual first offenders, and that the two sides were fairly equally attacked. True Chorea confined to the face is not common, though the disease often lingers here longer than elsewhere; the condition known as "Habit Spasm", or "Habit Chorea", is, on the contrary, very usually confined to the face, but is quite a different disease. Momentary diplopia from irregular movements of the eyeball muscles is an occasional symptom.

The tongue muscles and such muscles as are concerned in that part of deglutition which is voluntary, are/

are often affected by Chorea. Beevor* has called attention to a very characteristic smacking sound produced by the tongue being pressed against the hard palate and then suddenly withdrawn. The involuntary movements extend also to the trunk muscles and to the muscles connected with respiration, thus one often sees cases where the respiratory movements are markedly irregular, often giving rise to curious grunting expiratory sounds.

Von Ziemssen** describes various irregular movements of the laryngeal muscles, which can be seen by laryngoscopic examination, and this, I have no doubt, is a perfectly accurate observation, though so far as I know, it has never been confirmed. The difficulties of laryngoscopy in a choreic child can only be appreciated by one who has attempted it.

Chorea does not appear to involve the muscles of organic life. Patients do not suffer from colicky pains in the abdomen, nor from involuntary actions of the rectum or bladder. The irregularity of the pulse sometimes present can be explained on other grounds than cardiac chorea.

Braxton Hicks reported a case of Chorea during pregnancy/

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* Diseases of the Nervous System, Beevor, p. 330.

** Von Ziemssen, loc. cit., p. 436.

pregnancy, where at labour, irregular movements of the uterus were noticed, but reasoning on the analogy of absence of Chorea from other involuntary muscles, I should regard this as doubtful. It would be quite easy to mistake irregular contractions of the recti for those in the uterus.

The influence of voluntary movement is usually to increase the choreic but in some cases the reverse is the case. This latter effect is more likely to be found in cases of hysterical chorea than in the ordinary form.

Slight degrees of Chorea can often be brought out by asking the child to hold out both its hands horizontally in front of it and at the same time to put out its tongue, or to do any similar co-ordinated act. Excitement, mental or physical, is apt to increase the chorea markedly.

2. Muscular Weakness:

More or less weakness of one or more limbs or of certain muscle groups is not uncommon and occasionally the weakness is so great as to merit the title of Paralysis. Gowers* especially has drawn attention to this "Paralytic Chorea" and goes so far/

* Gowers, loc. cit., paragraph on Diagnosis of Chorea.

far as to say that "as a rule, when a child between 7 and 12 years of age is said to have gradually lost the use of one arm, the disease is Chorea." The age of onset and its gradual nature are the distinctive features.

The paralysis may be monoplegic, hemiplegic, or diplegic, but is much more commonly the first and confined to the arm. It may precede the development of choreic movement and thus may give rise to some difficulty in diagnosis.* Clifford Allbutt mentions a case of a boy whom he diagnosed to be suffering from Chorea merely from his having a paralysis of one finger; this diagnosis was confirmed by the patient developing choreic movements a few days later.

The paralysis is usually towards the end of an attack and is not often of long duration, though cases lasting months or even years, have been recorded. When seen in girls about the time of puberty, it may be mistaken for hysterical paralysis.

When hemiplegic in character, one may be in doubt as to whether the condition is true Chorea or the so-called post-hemiplegic Chorea. A case where this difficulty arose occurred in my series:-

D.B./

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* System of Medicine, Vol. 7; Article, Chorea by Russell, foot note by T. C. Allbutt.

D.B., Male, age 9-3/12th years. Admitted December 30th, 1902.

Complaint: Chorea of left side, mental backwardness.

History: His father had died of Phthisis and the family were in very poor circumstances. At the age of two, patient had been in Pendlebury Hospital with left hemiplegia, the cause of which was unknown. He had been in hospital three or four times since with clonic movements of left arm and leg. He had had measles twice, but otherwise his previous health had been good. He had never had Rheumatism in any form.

On Examination: He was found to have marked and incessant choreic movements of the left arm and leg, and to a less extent of the face, but here the affection was bilateral. There was very great weakness in the left arm and leg and also incoordination. He was able to stand and walk a little, but only with support. The knee jerks were both much exaggerated, especially the left. No ankle clonus. The plantar reflex gave a doubtful Babinski's sign on the left side. He was very backward mentally and spoke little, this being apparently partly due to the choreic movements of the lips and tongue and partly to his mental condition. His/

His cardiac condition was normal. He was treated at first with Hyoscine and later with Bromide, but did not improve at all.

On January 17th, the temperature rose to 100° and patient complained of headache and sore throat. The throat was red, but no membrane was present. The temperature remained between 100° and 102° for five or six days; the throat still continued red, but no membrane appeared and a swab taken, proved to contain no Diphtheria bacilli. No rash appeared. On January 24th, faint roughening of the mitral 1st sound became audible, which later, increased to a definite bruit. His urine was now albuminous and he had some diarrhoea. He was put on Sodium Salicylate and his temperature gradually fell to normal by February 2nd. The murmur remained at the apex. The mitral 2nd sound was reduplicated, and the second at the base loudly accentuated. He was discharged on March 5th, with only slight improvement. This case is interesting, as at first it appeared from the history to be more likely one of post hemiplegic chorea depending on some gross cerebral lesion. The attack of tonsillitis and endocarditis during the patient's stay in hospital appeared, however, to be of undoubtedly rheumatic/

rheumatic nature, so that the case seems to have been a somewhat unusual one of true hemiplegic Chorea.

3. The third point to be considered is Inco-ordination. This is practically always present to some extent at least, though sometimes it is difficult to separate from the results of interposed choreic clonic movements interrupting the performance of a motor act.

It may be very marked in cases where the general chorea is slight and may be found on the "healthy" side in a case of hemiplegic chorea.

Very interesting observations on the power of writing have been carried out mainly, in this country, by Russell* and in America by Weir Mitchell and Rhein.

Dr Russell divided cases into five groups according to their power of writing, these are:-

1. Cases with little or no control over the movements, inco-ordination probably present in most cases.
2. Power of control practically complete, no inco-ordination.
3. No choreiform movements or only very slight, much inco-ordination.

4./

* "Lancet", April 1, 1899. "Motor Phenomena of Chorea", J. W. Russell.

4. Control over chorea perfect but much incoordination.

5. Movements moderate, much central defect, writing impossible.

The first of those forms is the commonest, as one would expect. The second may occur even in severe cases with much choreic movement and is not common. The third variety may be seen, for example, in a case of left-sided Chorea, where there is no choreic movement in the right hand, and yet writing is very bad. This form is of great interest from a pathological point of view. The fourth is also rare and shows well the disorder of co-ordination, which may be so marked in Chorea.

The last group is that in which, apparently from mental involvement, writing is impossible, and in it too, speech is usually similarly affected. The movements nearly always cease during sleep, though that may often be difficult to obtain. The significance of this as indicating the site of the morbid process, is obvious.

Other disorders of the motor system are unimportant. In a few cases, spasm has been present, and in one or more of the limbs there has been a tonic contraction replacing the usual clonic ones.

In/

In a few instances, convulsive attacks of an epileptiform nature have occurred, and one of Gower's cases degenerated into chronic Epilepsy.

The Electric Reactions of the Choreic muscles is best studied in cases of hemichorea. Very often the A.C.C. is equal to, instead of being, as is customary, less than the C.C.C.

Sensory System: Chorea is usually a painless disease; sometimes vague pains in the muscles are complained of. Headaches are occasionally a source of trouble. Sensibility to touch is usually unaltered, though slight dulling has been noticed in a few cases, more especially in those with a hysterical element.

Mental Symptoms: The mental changes in Chorea are amongst the most interesting and instructive. In many cases there is throughout, no impairment, but in many others, this is present to a more or less marked extent.

Often the mental changes are very early symptoms; thus the child is peevish, irritable, and forgetful. She is unable to concentrate her attention on her lessons and is punished in consequence, and as Sturges* says:- "Infirmity provokes punishment, and punishment aggravates infirmity."

The/

* Sturges, loc. cit., p. 81.

The severity of the mental symptoms is not necessarily in correspondence with that of the bodily ones.

A choreic child has often a markedly stupid appearance, partly due, no doubt, to loss of tone in the muscles of expression, but also to the condition of mental dulness.

The more severe forms of mental disturbance, constituting "Chorea Insaniens" are not common, and when they do occur, are especially apt to do so rather later in life than usual, or in pregnancy. There may be very violent mania, or only hallucinations and delusions.

Speech: Interference with speech is seen in a large percentage of cases, and varies in degree. There may be only a slight hesitancy or precipitateness in the delivery of some words, or the interference may be so great that the child is quite dumb for the time being. In the majority of cases, the speech defect is due to inco-ordination in the mechanism of articulation, for the child can in most cases phonate, but in a few there is evidently interference with the laryngeal and thoracic muscles and in these, phonation may be impossible or impaired. In others, it may be in part due to the/

the patient's enfeebled mental state, and in still others, it is possible that it may be due to direct involvement of the cortical speech centres.

Reflexes: These are rarely altered. I have sometimes seen the superficial skin reflexes rather brisker than is usual even in children. The plantar reflex is always of the flexor type. Collier* in his investigations into this point, never found an extensor response in a case of Chorea. The deep reflexes are usually normal, sometimes the so-called "hanging up" of the knee jerk is seen, i.e., the extensor tendon remains contracted for a second or so longer than usual, so that the leg is held up. In my experience it is a rare phenomenon.

Temperature in Chorea: In an ordinary case there is little alteration in the temperature. Any great rise is in most cases a sign of endocarditis, or rheumatism, and should be looked on with suspicion.

In maniacal Chorea the temperature is usually high and hyperpyrexia may be seen, but hyperpyrexia may occur in an ordinary case and is an occasional cause of death. This is of interest as showing the alliance to Rheumatism. Cases have been recorded by several writers, e.g., Carpenter.**

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* Brain, Vol. 85, "An Investigation upon the Plantar Reflex," James Collier.

** Brit. Med. Jour., Aug. 29, 1903, p.454, Carpenter.

Affections of the Circulatory System:

These are of the greatest importance. In a very large number of cases, cardiac murmurs are present at one time or other in the course of the disease, and many theories as to their causation have been advanced.

Taking the actual clinical facts first, we may note that murmurs usually systolic in time and more especially heardover the mitral area, are present in a certain proportion of cases. As a rule there are no subjective symptoms complained of, though in some cases, there may be slight precordial distress, quickened action of the heart, sometimes palpitation, or dyspnoea, and slight rise of temperature. The physical signs are usually, a slight increase in the area of cardiac dulness, perhaps a more diffused and feebler apex beat than normal, with blurring of the mitral 1st sound, or an actual systolic bruit. The second sound at the base in the pulmonary area may be accentuated, but it must be remembered that in young children, the pulmonary 2nd is normally louder than the aortic.

Other murmurs are rarer, but may occur. The ordinary bruits of anaemia are often present in anaemic patients. Pericardial friction is occasionally/

ionally heard. The pulse in chorea is often quickened, and may be irregular apart from any sign of cardiac disease and in such cases this is often due to irregular respiratory rhythm.

In some cases we can study the course of the cardiac murmur from its origin to its termination. For example, it is common to discover in the first or second week of a mild attack, that there is a faint mitral systolic bruit; this becomes more audible, persists for some weeks, and ultimately grows fainter and disappears just as a similar bruit may in a case of Acute Rheumatism. But in other cases, the murmur remains permanently, or at least, for as long as the patient is under notice, and other definite signs of organic heart disease may develop, such as distinct increase of cardiac dulness, accentuation of pulmonary second sound, and perhaps signs of embarrassment of the right heart with its appropriate symptoms.

Now, there can be no doubt as to the nature of the change in the latter of those two groups, there has been mitral endocarditis followed by shrinking of the valve segments leading to incompetence of the valve, but it is with regard to the first group that opinions differ. We may arrange the various theories in four groups.

1./

1. Anaemic Theory:

It has been argued that the murmurs are due to anaemia, and in some cases no doubt they are, but against this, we have the fact that though anaemia is present in a large number of cases of Chorea exhibiting those symptoms, it is by no manner of means present in all. Besides, the commonest murmur is mitral, and not basal, as is usually the case in anaemia.

2. Another and more ingenious, if less correct view, is that the murmur is due to irregular contractions of the chordae tendineae of the mitral valve; in fact, that the Chorea has extended to the muscular apparatus of the valve. As Gowers says, "This is a view that can neither be proved nor disproved," but nevertheless there are strong arguments against it. Its supporters urge in its favour that there is no, or very little, increase in the area of cardiac dulness, and that the murmur is not present with every beat of the heart; but against this, we have the fact that the murmur varies little in character (one would expect its pitch and duration to vary greatly if due to irregular contractions); the murmur too, may be present in very mild cases and/

and absent in very severe ones. Besides, we can hardly imagine the Chorea to be confined to the musculi papillares of the chordae and not to spread to the ventricular well, and though the pulse is not uncommonly irregular, there is no evidence that this is due to Chorea of the ventricular wall. The arguments from the analogy of the absence of Chorea from the other involuntary muscles, are also of importance.

3. Another explanation is that the choreic movements lead to alterations in blood pressure, which cause a murmur, but why especially a mitral murmur, and why may it be present even in mild cases and not always in severe cases?

4. The view that is now most generally held is that the murmur is in the large majority of cases due to slight dilatation of the heart from myocardial weakness due to toxic action, or to myocarditis, or to endocarditis, or to a combination of those factors. Clinically to distinguish between those conditions in an early stage is an impossibility and probably they are usually present together to some extent.

As will be mentioned later, endocarditis is found in an enormous proportion of all cases dying of/

of or with Chorea. It is, as a rule, confined to the mitral valve and is shown by the occurrence of a ring of minute, friable vegetations on the auricular surface of the valve. How these tiny vegetations should be able to cause a murmur, is difficult to understand and possibly an associated dilatation is the chief agent in its production, for it is a well known fact that a murmur is not invariably present in cases with endocarditis. Myocarditis and an unusually diffuse fatty degeneration of the myocardium are fairly frequently found post mortem.*

As to the cause of this myocarditis and endocarditis, it is a very striking fact that cardiac affections in Chorea are more common in patients with a Rheumatic history, than in those without.

In the Pendlebury series, the following are the figures. I have classed them under the headings of cases with undoubted organic disease; cases with doubtful organic disease (i.e., cases presenting, for example, a bruit which only lasted a short time, or an impure mitral first sound, etc.), and cases with no cardiac lesion.

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* Fisher, British Medical Journal, August 29, 1903, p. 453.

	With definite Heart disease	With doubtful	With none.
Cases with history of Rheumatism. 27	8 = 29.6%	9 = 33.3%	10 = 37%
Cases without history of Rheumatism 38	2 = 5.2%	18 = 47.3%	18 = 47.3%

From this it will be seen that more especially in the cases with evident organic disease, there is a greatly increased percentage of such in those with a history of previous Rheumatism. Gowers says that heart disease is present in three quarters of the cases either before or during the attack of Chorea in those with a Rheumatic history, and in only one third of those without.

The statistics of the British Medical Association Collective Investigation give 50% of heart disease in the rheumatic cases, and 35% in the non-rheumatic.*

The most satisfactory explanation of the occurrence of myo- and endocarditis in Chorea is that Chorea is an infective disease and that the cardiac involvement is due either to the direct influence of the organism of the disease, or to the action of its toxins. The comparatively recent researches of Walker and Beaton, and Poynton and Paine and others have still further strengthened this view. These will/

* B.M.J., Feb. 26, 1887, p. 432.

will be referred to later. As analogy, we have the cardiac involvement of Rheumatism and the close relation of that disease to Chorea and its probable infective nature is undoubted. We have also the analogy to Pneumonia, Gonorrhoea, Septicaemia, etc., which are all diseases where organisms and their products are the cause of the endocarditis. The different views on the pathology of Chorea will be be discussed later, but we must mention here that the supporters of the "functional" nature of the disease find this fact of the occurrence of Endocarditis somewhat difficult to explain; while the advocates of the "embolic theory" have been so struck with the cardiac features of the disease that they have set up endocarditis and embolism as the cause, rather than a result, quite forgetting to go to morbid anatomy as a guide and apparently unaware that endocarditis is by no means always present, and that if so, their theory becomes untenable.

Morbid Anatomy:

Statistics of fatal cases have been collected by Sturges, Raymond, and Osler.

Sturges* found that in 80 cases, no fewer than 75 showed endocarditis or pericarditis.

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* Sturges, loc. cit.

Raymond* in 19 cases found endocartidis in all.

Osler** in 73 cases found endocarditis in 62. Thus in a total of 172 fatal cases collected by those observers, endo- or pericarditis was found in 156, a percentage of 90.7%. In no other fatal disease is endocarditis so commonly found post mortem as in this. In acute Rheumatism, in a series of 45 fatal cases collected by Fagge,*** in 19 endo- and pericarditis were present together, pericarditis alone in 10, endocarditis alone in 8. The number of cases where heart unaffected was 8, or a percentage of 17.7%, whereas in the cases of Chorea, the percentage where heart unaffected is only 9.3%

The endocarditis is usually of the simple variety and is especially prone to affect the mitral valve. On examining its auricular surface, one finds a ring of tiny vegetations, which are at first soft and friable, but later, get firmer and denser. The changes as seen microscopically are thus described by Poynton.*=*

1. Swelling of subendothelial tissue,
2. Cellular proliferations,
3. Necrosis of portion of valve.

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* Raymond quoted by Osler in monograph "On Chorea", p. 51.

** Osler, "On Chorea", p. 53.

*** Fagge, Practice of Medicine, Vol. I., Acute Rheumatism.

= "International Clinics", Vol. II., Series XIII.

A cap of fibrin is not an essential, though it is a usual part of a vegetation. He has been able to isolate from the affected valves a diplococcus which is identical with that of acute Rheumatism.

The other valves are not so often affected, probably the small blood vessels in the mitral and its more complex structure, render it more easily attacked.

Gowers* only found two cases of pure aortic disease in 250 cases. The British Medical Association Collective Investigation Report,** gives six pure aortic cases in a total of 135, but this, I think, is an abnormally high proportion. More than one valve is occasionally affected.

Occasionally the endocarditis is of the ulcerative variety and embolism may follow. Several examples of embolism of the Arteria Centralis Retinae have been recorded. Thomas of Baltimore*** has collected seven cases, including one that came under his own personal notice. Similarly, emboli in other organs are occasionally met with, e.g., in the kidney.

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* Gowers, loc cit.

** B.M.J., Feb. 26, 1887, p. 431.

*** Johns Hopkins Hospital Bulletin, October 1901, p. 321.

Pericarditis in Chorea is not very common. In my own 65 cases, I met with no instance of it.

Osler found in his 73 fatal cases pericarditis in 19, in 17 of which it was associated with endocarditis.

Myocarditis: Cardiac dilatation is very common and is probably in most cases due to toxaemic weakening of the Myocardium, but true myocarditis is sometimes found. Fisher* found in one case marked fatty degeneration, the distribution of the granules being rather different from that usually seen.

Post mortem evidence is thus a strong support to the view that the heart murmurs of Chorea are of an organic nature, for there is no reason to suppose that the cardiac condition in a fatal case is in any material manner different from that in a non-fatal.

Sturges has attempted to show that the endocarditis of Chorea is of two types:-

1. Choreic,
2. Rheumatic, occurring in a choreic child.

He has endeavoured to make out that the former is entirely benign, harmless and temporary. Thus, on/

* B.M.J., Aug. 29, 1903, p. 453.

on p. 70 of his monograph, he says: "Those later changes leading to valve distortion and consecutive alterations in the heart's chambers, which belong to Rheumatic endocarditis, have no place at all in that which belongs to Chorea."

The after history of such cases, however, proves only too clearly how very far from true this is.

STATISTICS OF THE FREQUENCY OF HEART LESIONS:

Statistics as to the frequency of heart murmurs in Chorea are very numerous, but very few writers have arranged their figures to show in how many those murmurs were due to organic lesion, in how many due to dilatation, and in how many to anaemia, etc., and to distinguish between those, if possible, is of some importance.

I have endeavoured to allot my cases to three groups. In the first are placed cases where the physical signs are obviously due to organic disease; thus, all cases are included here which had undoubted murmurs, especially if rough or high pitched, where the murmur persisted, also cases with much enlargement of the heart, especially if there seemed to be cardiac hypertrophy as well as dilatation.

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In the second class, I include cases which have presented at one time or another a murmur usually faint, or where the mitral 1st sound was merely short and impure, and where this murmur disappeared under treatment; also cases of cardiac dilatation as shown by feeble pulse and apex beat with some enlargement of the heart. This latter group may present a murmur, but not invariably. In many of those cases, there may be endocarditis, but I have thought it wiser to include them in this so-called "doubtful" category.

My results were as follows:-

Total	Cases with Organic Lesion	with "doubtful" Organic Lesion.	without lesion.
65	10 = 15.3%	27 = 41.5%	28 = 43%

If we add the 1st and 2nd groups together, the percentage of cases which had at one time or other some evidence of cardiac derangement is 56.8%.

I have summed up the statistics of the British Medical Association Collective Investigation, of Ashby*, Osler, Sturges, and of Fagge with the following result:-

Out of 1979 cases, heart murmurs or irregularity was present in 765, a percentage of 38.6%.

We have next to consider the influence of recurring/

* Diseases of Children, Ashby and Wright, 4th Edit., p. 519.

curring attacks, or the incidence of heart affection, and this undoubtedly is to increase the liability.

The following are the statistics of my own cases:-

	Definite Heart Lesion	Doubtful	None
1st Attacks (36.)	3 = 8.3%	13 = 36.1%	20 = 55.5%
2nd " (21.)	4 = 19.04%	11 = 52.3%	6 = 28.5%
3rd or more (8.)	3 = 37.5%	3 = 37.5%	2 = 25%

This gradual rise with successive attacks may be explained in three ways:-

1. The endocarditis may be due directly to the Chorea organism, or its toxins and therefore each successive attack exposes the heart to a fresh risk.

2. With each year after an attack of Chorea we get an increase in the number of cases which suffer from Rheumatism and thus we have the heart exposed to fresh risk from this cause.

3. Osler followed out the after history of a large number of cases and found that an enormous proportion after some years, showed organic heart disease even though they had not had fresh attacks of Chorea, or Rheumatism, and therefore when we examine the heart in second or later attacks, there has been time for previously latent lesions to become developed.

Probably all of those factors are at work.

AFTER HISTORY OF CASES OF CHOREA WITH REFERENCE TO HEART DISEASE.

Stephen Mackenzie and Donkin are quoted by Osler* as having examined cases with a view to determining this point. Mackenzie in 33 cases examined at a period of from one to five years after an attack, found organic heart disease in 60.6%.

Donkin, in 44 cases examined at from 2 to 12 years after an attack, found 18 or 40.9% with heart disease. Osler in his own very careful investigation of 140 cases at a period of from 2 to 16 years after the attack of Chorea, found that 72 cases or 51-3/7% had definite damage to the heart, in 17 cases there was at least functional disorder, and in 51 the organ was healthy.

In the 72 cases with organic disease, 30 had had 3 or more attacks of Chorea, and in 25 there was a history of acute arthritis which in 7 had followed the Chorea in from one to five years. In this group of 72 cases thus the percentage with a history of acute arthritis was 34.13%; in those 51 cases without heart lesion, it was only 17.11%, showing very clearly/

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* Osler, loc. cit., p. 54.

clearly the importance of rheumatism in the etiology of valvular disease. But 66% of the 72 cases which had developed heart disease, gave no history of rheumatism in the interval.

Fisher* gives statistics of 53 cases which he examined not less than three years after the attack of Chorea. He found definite organic disease in 37.7%. He mentions also having seen cases where no organic disease, nor dilatation could be detected, yet the patient complained of inability for severe exertion, these cases no doubt having some myocardial damage.

To sum up, we may say that nearly one out of every two children who have Chorea will suffer from cardiac disease in later life.

I examined 38 cases of organic heart disease (excluding congenital cases) in Pendlebury Hospital in 1902, to find in what proportion of cases Chorea had been an etiological factor. It was found in 5 cases, a percentage of 13%; in 4 of these there was a history of arthritis as well.

The usual lesion found in later life due to choreic endocarditis is mitral incompetence; mitral stenosis is considerably less common, and so also are aortic lesions.

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* Fisher, loc. cit.

AFFECTIONS OF OTHER SYSTEMS:

These are but rarely affected. In the respiratory, we may have Pneumonia and Pleurisy, possibly an indication of an unusually severe and virulent infection.

In the Urinary System: The urine is often of rather high specific gravity; phosphates and urea are said to be increased. Nephritis is a rare complication. Albuminuria and haematuria may, in rare cases, occur as the result of renal infarction.

Cutaneous: Skin rashes are not uncommon. Herpes Zoster is occasionally seen, apart from the administration of Arsenic. Rheumatic eruptions, as Erythema Nodosum, are not common. Naturally, arsenical eruptions from the common use of this drug in large doses, are often met with. Of these, the commonest, are Herpes Zoster,(of which I have seen two cases,)Erythema and Pigmentation.

VARIETIES OF CHOREA:

For convenience' sake, we may describe three grades of severity, - mild, severe, and maniacal.

Mild: Fortunately this is the commonest variety. It is characterised by not very exaggerated movements, little affection of speech, and as a rule by no/

no very marked mental deterioration. It is often surprising how little the patient, especially the younger ones, seem to mind their infirmity.

Severe: In this, the movements are severe, the speech often lost, and there is more likely to be mental deterioration. The movements may be incessant, and widespread, and the child is quite unable, in most cases, to perform any voluntary act. Great difficulty may be experienced in the feeding of the patient and I have known of a child, who, when being fed from a cup, managed to bite a very respectable piece out of it. Sleep, from the violence of the movements, may be impossible.

Maniacal Chorea: This is fortunately a rare form, for it is a very terrible one. A mild case of Chorea may be converted into this form by injudicious exposure or treatment. True Chorea Insaniens is characterised by very violent mental symptoms, such as acute mania, hallucinations or delusions. The temperature is often high and there amy even be hyperpyrexia. Sometimes the choreic movements may be so slight as to be overlooked and the patient may be sent to an asylum as an ordinary case of insanity. This variety of the disease is oftenest met with in pregnant women and in girls about/

about the time of puberty. There is usually very rapid emaciation, and death is by no means infrequent.

Other sub-varieties of Chorea are chronic and senile.

Chronic: Chorea, though usually a disease of a few weeks' duration, may in a few cases become chronic. Cases where it has lasted for years and even for life have been recorded by several reliable authorities. Such cases are more apt to occur in adults.

Senile: Senile Chorea is in many instances an entirely different disease from what we have been describing. In some cases, it is probably allied to Huntingdon's Chorea, but in a few, it appears to be a true Chorea of Sydenham. Thus, cases where the patient had suffered from Chorea Minor in childhood and where the course, etc., of the attack in old age were identical, have been described.

Cardiac affections and rheumatism are said to be relatively uncommon. Bischoff* gives only 12% of cases with heart lesion.

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* Quoted in American Yeer-book of Medicine, Gould, Vol. I., 1903, p. 431.

DIAGNOSIS:

No disease can, as a rule, be more readily diagnosed than Chorea. The character of the movements, the mental condition, the involvement of speech, the cardiac affection, the history, age, and sex of the patient are all factors that help us in our diagnosis.

Cases in which difficulty arises do occur, however. Of these, we may mention, first, cases of Paralytic Chorea. This rarely causes any trouble in diagnosis. The paralysis is not often severe, and besides, usually succeeds the choreic movements. In cases where there has been no chorea, one would rely on the facts of its somewhat gradual onset, that it is usually confined to one limb or to one side of the body (~~but not of the face~~), and that it is seen in a child of about the age at which Chorea is common. It has no resemblance in most cases, if a careful examination be made, to hemiplegia due to gross cerebral lesion, or to infantile paralysis.

Hysterical Cases: This type is not uncommon. One is usually able to find other evidence of Hysteria; the movements are rhythmical, and are very/

very markedly increased when the patient is aware of being watched. Cases where Chorea occurs in a hysterical subject, may give considerable trouble, and are pretty often met with. The following is a case illustrating this connection:-

Gertrude R ----, a fairly well developed girl of 11, was admitted to Pendlebury Hospital on August 6th, 1903. Her home surroundings were poor, her father was out of work and there were eight other young children. Her previous health had been fair. She had had measles in infancy. There was a vague history of "growing pains". At the age of seven, she had an attack of Chorea, for which she was treated as an out-patient for eleven weeks. She had a second attack six months later. Her present illness began a fortnight prior to admission, with headaches and photophobia. A day or two later, she developed Chorea.

On Admission the Chorea was violent and constant. Her speech was a little jerky. She was an intelligent child, and of a distinctly neurotic type. There was a faint mitral bruit and slight cardiac dilatation; during her stay in hospital this gradually passed away.

She was treated at first with Bromide of Potassium/

assium gr. 20 and Chloral Hydrate gr. X T.I.D. On the evening of August 15th, the Chorea became extremely violent and patient was quite delirious, but throughout the delirium she was unusually well able to recognise the sister and nurses and other patients, and often made exceedingly á propos remarks concerning them. She also had opisthotonos. She was given morphine hypodermically and later, inhalations of Chloroform and became much quieter. A few days later, her Chorea was very slight, but patient suddenly began to vomit every thing she was given, even peptonised milk. This lasted a few days and very rapidly ceased after she was isolated in a special ward. A few days after this, the Chorea again became severe and patient became noisy, but seemed to improve after she was put on Antipyrine gr. X. every four hours. She was ultimately discharged quite well on September 20th.

She was readmitted with another attack a month later and the course of this was very similar. It was noticed very markedly how the choreic movements became exaggerated when she was aware of being observed.

Motor Tics: This condition of Motor Tic or Habit Spasm, is one in which some one or more involuntary movement/

movement is repeated over and over again. It is found in nervous children about the time of puberty. The movements are commonest about the face, and are such as blinking of the eyes, sniffing, etc. These movements often have a reflex origin, e.g., blinking from eye defects, sniffing from adenoids or nasal catarrh, and so on.* The movement is begun as a "trick" and becomes involuntary and beyond control. It is sometimes a little difficult to distinguish a purely facial chorea from this condition of habit spasm. The history and progress of the case will help one in deciding. The movements in habit spasm too, are more localised, neither is there any similar mental condition, heart lesion, or speech defect as a rule.

Pre- and Post Hemiplegic Chorea:

These conditions are practically of little importance in the light we are now studying them. The movements are of an entirely different type in the majority of cases, and there are many other distinguishing features.

Electric Chorea: This is a disease first described by Dubini about half a century ago. It is an/

* See Allchin's Manual of Medicine, Vol. III., 1901, p. 348.

an infective disease mainly located in Italy. It has derived its name from the shock-like nature of the contractions from which the patient suffers. It is fatal in 90% of cases and in this country, at least, can not be any source of difficulty in the diagnosis of ordinary cases of Chorea.

Huntingdon's Chorea: A disease of adult life with a strong tendency to hereditary transmission. The movements are slower than those of Chorea Minor. It precedes gradually to mental enfeeblement and death. Pathologically, it is a chronic meningo-encephalitis.

Hereditary Ataxia and Paramyoclonus Multiplex:

Bear only a very superficial resemblance to Chorea and it is unnecessary to consider them.

COURSE:

The duration of cases varies greatly, probably from six to ten weeks is the average. In the Pendlebury series, the average stay in hospital was 47 days or about 7 weeks. This, of course, does not give total duration, as most cases had had Chorea for from some days to some weeks before admission./

mission. Each case too, was usually kept in hospital for a week after complete cessation of movements.

Very acute cases occasionally occur, and cases of Chorea Insaniens may be fatal within a week. Occasionally, the disease becomes very chronic and may last for months, or years. Gowers mentions a case which lasted from youth till death, at the age of sixty-six.

Protracted cases are commonest in boys. In my own series, the average duration in boys was 51½ days, compared with 42 days in girls; the longest cases, however, in this series, namely 102 and 107 days, were in girls.

The severity of the attack is no guide as to its probable duration. Gowers was unable to "discover any relation between duration and age, sex, state of heart, preceding Rheumatic fever, or exciting cause."

Once improvement has begun, it is as a rule eapid; the Chorea soon disappears, but may be often reawakened by excitement or emotion. Recovery is usually perfect, but sometimes some inco-ordination, needless haste in executing certain movements, or facial grimaces, etc., remain for years, or even permanently.*

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* Von Ziemssen, loc cit., p. 444.

MORTALITY:

Is usually given as 2% (9 out of 439 cases, B.M.A. Collect: Invest: Report.) In the 65 cases which I have examined, and in 554 cases attending the Manchester Children's Hospital Out-Patient Department during 1901-1902, a total of 619 cases, there was no fatal case.

Death is commonest in girls about puberty, or during pregnancy. It may result from exhaustion as in very severe cases, from heart complications especially pericarditis, from hyperpyrexia, from Pleurisy or Pneumonia, etc. Cerebral haemorrhage has been an occasional cause; so also has Pyaemia following injuries sustained during the constant movements. Fagge* mentions a case where fatal obstruction to respiration was produced by a glossitis due to injury to the tongue. A death from acute dilatation of the stomach has been described.

The especial frequency of death in cases of Chorea Gravidarum is often due to labour or abortion in an exhausted patient.

RECURRENCES:

These are very common, and it is usually well to/

* Fagge, loc. cit., p. 884.

to warn parents on this point. Recurrence is apt to occur within two years of the previous attack; after this time, the danger is greatly lessened. American writers have laid stress on the tendency of recurrent cases to occur in spring, but in this country, there does not appear to be any such tendency. Weir Mitchell* mentions a case where a girl had Chorea four times in March or April, then in February, and then twice in May. He states that in 25 recurrent cases, 19 recurred in spring. Gowers describes the case of a girl who had nine attacks, most of which were in autumn.

PENDLEBURY SERIES:

	1st Attacks	2nd Attacks	3rd or more
Boys	8	4	2 = 14
Girls	26	17	6 = 49
Total	34	21	8 = 63

It will be seen from this table that 29 cases had more than one attack, a percentage of 46. and 8 cases more than two attacks, a percentage of 12.7

The course of a recurrent attack is, as a general rule, very similar to that of a first, its duration often being a little less. The figures for/

* Diseases of Nervous System, S. Weir Mitchell, p. 147, (2nd Edit.)

for my cases are as follows:-

1st attacks ..... 49½ days approximately,
2nd " ..... 45 " "
3rd or more ..... 45 " "

Sée's results as quoted by Church* are 139 days, 80 days, and 55 days, which gives a very marked shortening with each successive attack, but I can not help thinking that an average duration of nearly 20 weeks for a first attack is unusual.

With each succeeding attack, one is more likely to get cardiac involvement, though some writers have stated that if the heart escape in the first attack, it is less likely to be attacked in subsequent ones. This, however, is incorrect.

MORBID ANATOMY:

The absence of any distinctive lesion has always been one of the stumbling blocks in the road to a proper view of the true nature of Chorea. This absence of distinctive lesion has given rise to the belief in the so-called "functional" nature of the disease, a view which, besides being erroneous, explains nothing. The only distinct and marked lesion found in fatal cases, is endocarditis, which is/

* Church and Paterson, Loc. cit., p. 538.

is found in something like 90% of cases, but endocarditis and the rarer post mortem findings, such as pericarditis, pneumonia, pleurisy, nephritis, etc., have no direct bearing on the production of the Chorea itself.

In the nervous system to which we naturally turn for some physical basis for the disordered action of the muscles, we find very little indeed.

On a naked eye examination one usually finds decided cerebral congestion, minute haemorrhages and softenings, and sometimes emboli. One or two cases of definite apoplexy have been reported.

Microscopic Examination: Here also one finds nothing distinctive, though of course, the brain is, as Poynton says, an enormous organ to investigate satisfactorily with 1/12 objective. There are the same signs of cerebral congestion, dilated perivascular spaces with some accumulation of leucocytes, here and there patches of softening or minute haemorrhages, and occasionally, small emboli or thrombi.

With regard to the nerve cells, often there is no abnormality, but sometimes there are the common degenerative changes found in many diseases of the nervous system - swelling and cloudiness of the protoplasm/

protoplasm, slight vacuolation, etc.

These changes are widespread through the brain, and to a less extent and especially in chronic cases in the spinal cord. There is not, as a rule, any special limitation to the cortex, though one observer (Turner*) found degeneration changes especially affecting the region of the deeper pyramidal cells in the Rolandic area. Out of the five cases, however, in which he found these changes, in two, there had been albuminuria and in one puerperal septicaemia.

Dana** in 1890 collected a series of 39 cases in which an adequate description of the morbid anatomy of the brain and cord was given. The above conditions were found by most of the observers, and in subacute cases they appeared to have been slightly more marked in the deeper parts of the motor tract, and in the lenticular nuclei and inner parts of the thalami. In the more chronic cases, the vascular and neuro-degenerative changes were more marked. The small arteries were more dilated, thickened, and degenerated and the perivascular channels more eroded and distended. There was also some connective tissue proliferation and signs of degeneration in the ganglion cells. These also were the changes found in two cases examined by the most/

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* Turner, Path. Soc. Trans. 1892, quoted by Church and Paterson, loc. cit., p. 533.

** "Brain", Spring number, 1890, p. 76.

most recent methods by Reichardt.*

In the cerebellum, no special changes have been described. There may be minute haemorrhages, etc., in the pons and medulla.

Spinal Cord: Here also we may have any of the above changes. Handford** in two cases of Chorea Insaniens examined post mortem by him, found small congestions and haemorrhages and a few thrombi mainly in the upper cord and pons, in addition to general cerebral congestion. Probably spinal cord changes are commonest in old standing cases and may be due to perverted function.

In Dana's own case, there was slight lepto-meningitis, but this was one of twelve years' duration.

Peripheral Nerves: Neuritis has occasionally been found even in cases where there was no suspicion of its being due to arsenic. Gowers, at the close of his description of the pathological anatomy of Chorea, says: "Such of the above changes as are not accidental concomitants of Chorea are probably the result of the excessive and perverted functional activity of the nerve elements, which is always attended by vascular dilatation in the part affected/

* Reichardt quoted in Amer. Year Book of Medicine, 1903.

** "Brain", July 1889, p. 129.

affected, and this may have its own consequences in thrombosis, extravasations, or leucocytal accumulations."

Besides this, the afore-mentioned conditions may be found in death from any acute infection, so that it is obvious that the morbid anatomy of Chorea is in no sense distinctive. Chorea in the dog is mainly a spinal cord disease, but is an entirely different disease from Chorea Minor.*

BACTERIOLOGY:

The view that Chorea is an infective disorder has long been advocated and the proofs of this are gradually being added to, and there is little doubt that they will before long be completed. Many different organisms, specific and otherwise, have been described in the heart valves and central nervous system in cases dying with Chorea, but up till lately, they have had to be set aside. One great fallacy in this putting forward of different bacteria is that the observer has neglected to take into consideration, the fact that during the last days of life a terminal infection may occur, which has nothing whatever to do with the original disease. A great deal of work has been done in the last few years/

* Lancet, Jan. 9, 1886, Horsley (Brown Lectures.)

years on the bacteriology of acute Rheumatism and we must consider this and see what bearing it has on that of Chorea. In 1891, Achalme discovered a bacillus in the blood of patients dying with Rheumatic Fever. He was able to cultivate it in several cases, but experimental innoculation in animals did not produce any condition resembling Acute Rheumatism. This bacillus was also investigated by Thiroloix, and this observer obtained some of the features of Acute Rheumatism on injection into rabbits. Subsequent researches have not, however, confirmed this organism's claims.

In 1898, Triboulet isolated a minute diplococcus in several successive cases and was able experimentally to produce endocarditis and pleurisy in rabbits, but not arthritis. This coccus has also been found by Westphal, Wasserman, Malkoff, and Mayer, and in this country by Paine and Poynton, and by Walker and Beaton. The organism is exceedingly minute, stains well with the ordinary aniline dyes, retains the stain by Gram's method, and has been cultivated. It does not agglomerate when treated with the serum of convalescent rabbits or man. Paine and Poynton* have isolated it from many successive cases of Rheumatism and often in pure/

* Lancet, Sept. 22, Sept. 29, 1900, May 6, 1901, and Path. Soc. Trans. (London), 1901.

pure culture. They have found it in the blood and urine of living patients, in the blood, heart valves, pericardium, joints, lungs and pleura, and kidneys in patients dying of Rheumatic Fever. Also in the pia mater and brain of a fatal case of Chorea, and in the throat of a patient with rheumatic tonsillitis. Innoculation experiments in rabbits have produced every feature of acute rheumatism in man, viz., arthritis, endocarditis and pericarditis, cardiac dilation, pneumonia and pleurisy, etc. The urine is acid and loaded with urates and there is moderate pyrexia. Anaemia is usually rapidly produced. Drs Paine and Poynton were not able to grow their organism on highly alkaline media, as had been done by Wasserman, but in other particulars, the organisms appear to be identical. Beaton and Walker* have fully confirmed the results gained by Paine and Poynton, and to some extent have extended them. They have worked out a very full table of cultural characters and have been able to grow the coccus on alkaline media. The coccus usually occurs in pairs or short chains and appears to be specifically distinct. Applying Marmorek's test, they were able to cultivate it in a medium in which pyogenic streptococci/

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* Brit. Med. Journal, Jan. 31, 1903.

cocci had been grown and from which these had been removed by filtration. This test is generally considered to show that the micrococcus Rheumaticus is specifically distinct, as ordinary streptococci will not grow in such a medium.

Walker and Ryffel* have investigated the chemical reactions of this organism. When cultivated, it is noticed that acid is produced rapidly. This acid has been found to be formic acid, and there is also formed, but to a lesser extent, another of the fatty acids. This acid is present both in the filtered cultures and in the bodies of the microbes themselves. It is believed to be produced by the oxidation of sarcolactic acid. They were also able to prove that formic acid was present in the urine and tissues of animals suffering from Rheumatic Fever and that its amount in the urine in man was diminished by treatment with Salicylates. Formic acid is not produced by ordinary streptococci.

Paine and Poynton, as has been mentioned, were able to demonstrate an identical coccus from the pia mater and brain of a fatal case of Chorea, as has also been done by several German observers and by Dana in America. They have also isolated the organism from choreic heart valves. The innoculation of the

* B.M.J., Sept. 19, 1903.

the coccus into rabbits produced , not Chorea, but Acute Rheumatism. In one of Poynton and Paine's rabbits, about three weeks after innoculation with the coccus, there was paresis of the hind limbs, which gradually got well. This, they suggest, as being possibly allied to a condition of paralytic Chorea. In a later case an innoculated rabbit became extremely nervous and manifested twitching movements of the limbs and face. Post mortem, they were able to show the diplococci in the pia mater and in the endothelial cells of the blood capillaries dipping into the motor cortex from the surface.* On another occasion, they isolated the organism from the cerebro-spinal fluid of a rabbit suffering from valvulitis and peridarditis, but showing no signs of Chorea. Beaton and Walker, though they innoculated several young rabbits with the diplococcus, were unable to produce any very definite Chorea. It must be remembered, however, that a certain type of brain cell is probably of great importance in the production of Chorea and it is not at all likely that an animal such as the rabbit should possess it.

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* Lancet, May 4, 1901, p. 1261.

PATHOLOGY:

In dealing with this part of the subject, it will be convenient to consider:-

1. The site of the lesion.
2. Its nature.

1. Site of the Lesion:

The movements in Chorea are, though involuntary, similar in character to the ordinary purposive movements of daily life, they are not movements of individual muscles.

Now, this fact points to a lesion in the upper neurone as distinguished from one in the lower. It is impossible to conceive of choreic movements being produced by lesions only in the nuclei of the motor cerebral nerves, or in the anterior cornual cells of the spinal cord for those cells are the source of the stimulus to contraction of certain definite muscles. There is some power of coordination in the cord, but in man only to a very secondary degree. The cells of the motor cortex are the initiatory factor in the production of any co-ordinated muscular act, thus we have the movement of say - flexing the arm presided over by certain cells in the/

the Rolandic area, and able to be produced by stimulation of that part, whereas the cells that are connected with the biceps muscle are seated in the spinal cord, namely in its 5th and 6th segments. The cessation of movements in sleep is in favour of a cerebral lesion, and considerably against one in the cord, for the reflex functions of the latter are known to be more active during sleep.

The occurrence of hemichorea also leads us to the conclusion that the site of the lesion is in the brain. It must be remembered too, that facial and trunk chorea is never unilateral, a fact most significant of a cortical origin, for those movements are represented in a different manner from those of say - the arm or leg. The distribution of the Chorea, the special liability for the face and hands to be affected is what we should expect, if the site were in the cortex, for these movements are represented in a relatively larger area than the less complex movements of the foot; movements are represented in the cortex by areas which are proportional in size to the complexity and not to the size of the muscles involved.

Now, those movements which are comparatively so lagrely represented in the motor cortex are also those/

those over which voluntary control is most complete. Thus, we have very much fuller control of facial, tongue, and speech movements than over those of the trunk and legs. In Chorea, those are the movements most prone to be affected, and if the lesion in Chorea be cortical, this is as we should expect. Similarly, we have more control over the respiratory movements than over those of the heart, over which, in fact, most of us have none at all, and so we not uncommonly get irregular respiration in Chorea, whereas irregular cardiac action, apart from cardiac disease, is uncommon. Certain of the movements connected indirectly with respiration are located in the cortex, whereas no cortical localisation of cardiac movements is found.

The mental involvement which is so exceedingly common, the slow and dull working of the mind, the fits of temper and of emotion, also point to a cortical lesion in another portion of the brain, and so to a certain extent, do the speech difficulties. As has been mentioned previously, speech is very frequently lost to a greater or less extent in Chorea and the explaination of this may be threefold.

1. It may be, and is most usually due to incoordination among the muscles whose delicate interworking is essential for proper speech. In most cases the articulative muscles are more at fault than the laryngeal or thoracic ones.

2./

2. The mental torpor so often present, may be so marked, that though able to perform properly, the muscular acts which go to the production of speech, the child makes no effort to do so.

3. Possibly in some cases the absence of speech may be due to direct involvement of the cortical centre (Broca's convolution) for motor speech. As Dr Lees* has shown this is practically the only explanation we can find for cases which have remained dumb for some weeks, or months after an attack from which otherwise they have perfectly recovered.
I have not been able to discover any record of investigations as to whether speech defect of this last type is ever found in cases of left sided hemichorea.

So far, therefore, all evidence points to the lesion being mainly in the brain, and to the cortex as being in all likelihood the chief portion involved, or to the lesion being at least either in the cortex or on the proximal side (if we may use such an expression) of it, in the production of a motor act. A voluntary motor act does not, however, strictly speaking, originate in the cortex; it must arise from those higher centres where what we call the "will" resides. Any voluntary movement is the result of a very complex process and one which is not fully understood, and it is necessary to say a few words on this subject here.

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* B.M.J., Aug. 29, 1903, P. 449.

It has been proved experimentally both in man and in the lower animals that stimulation of the Rolandic area of the brain leads to the production of co-ordinated movements mainly on the opposite side of the body, and also that the path of the nervous impulse is by means of the internal capsule and pyramidal tract to the motor cells of the nuclei of the cerebral nerves, and of the anterior cornua, and thence by the motor nerves to the muscles. But, besides this efferent tract from the motor cortex, there are also afferent impulses which are of no less importance in the production of a co-ordinated movement. The brain is continually receiving from the senses of sight, touch, hearing, from the muscular sense, from the semi-circular canals, etc., impulses which help in the co-ordination of movement. If those are wanting or interfered with, co-ordination is interefered with to a greater or less extent. How those separate impulses are blended, as it were, and what is the relative importance of each, is not at present known; some of them obtrude into the realm of consciousness, whereas others do not. No experiments of physiologists have as yet given us very definitely an area which we can look to as being the centre for the co-ordination of those afferent/

ferent impulses, but surely such an area or areas must exist, and so must it be connected directly or indirectly with the motor cortex and efferent tract. The optic thalamus has been supposed to be possibly a centre where some of those impulses, more especially those that do not awaken consciousness, are arranged.

Besides the impulses received from the senses, the motor cortex is also influenced by what is commonly called the "will". Thus, to sum up, - in the production of a co-ordinated motor act, we have on the one hand a number of impressions received from various senses; these are arranged and allowed for, possibly by some special centre. On the other hand, we have the cells in the Rolandic region and their efferent fibres which are able to conduct the required nervous impulses to the muscles to be used. Between those afferent and efferent systems, there lie the more complex arrangements of the brain known as the "intellect" and the "will".

In Chorea the change in the nervous system is widespread and there is probably some involvement of each of those factors, but in varying degress, and this more or less general involvement is in harmony with the view that the condition is mainly produced by toxic action on the nerve cells. The micrococcus/

micrococcus of Rheumatism has, however, been found in the pia mater and also in the grey matter of the cortex, so possibly its action is to some extent direct. Such facts as the occurrence now and again of hemichorea, etc., point towards the lesion being in part, at least, a focal one.

The higher centres of the brain, where the intellectual and volitional processes are carried out, are very obviously involved in a large number of cases. The sensory side of co-ordinated motion is probably less affected than the others, but very probably the difficulty in carrying out volitional efforts is in part due to this cause. The most obviously affected of all, is the efferent side, the choreic movements are such as we can easily reconcile with irregular and explosive working of the motor cells. There is no evidence of the chorea being produced by interference with the nervous impulses after their departure from the motor cortex; any interference must be either in the motor cortex or in those centres which influence it and whose activity in the production of a motor act occurs prior to that of the motor areas.

Lesions in the central ganglia, and more especially the optic thalamus, and in the crus cerebri may produce choreoid movements, but, as Gowers says/

says in his remarks on the pathology of Chorea, "It is doubtful whether there are at present any facts to justify us in going beyond the motor cortex in our search for the primary disturbance."

Dana* suggests that the phenomena of paralytic Chorea may be explained by the presence of some single large focus of congestion, exudation, or haemorrhage.

2. The Nature of the Lesion:

What the precise nature of the change in the nerve cells is, that should lead them to energise in so curious and intermittent a manner, it is impossible to conjecture. We have the analogy to a certain extent to epilepsy, to Huntingdon's Chorea, to the various Tics Convulsifs, all of which are diseased conditions characterised by clonic movements and most probably occasioned by lesions in the motor cells, but this analogy, though noteworthy as an indication of a psecial tendency of those cells in disease, helps us but little to understand its real nature. The lesion is in all probability an irritative one. The phenomena of Chorea in pregnancy have lead some writers to suggest/

* Textbook of Nervous Diseases, Dana, Edit. 4th, p. 501.

gest that in this form, at least, it may be a paresis of the inhibitory centres that is the cause, for it is known that in pregnancy, the powers of inhibition are weakened generally. This view would explain such facts as the frequency in first pregnancies and in illegitimate ones, for in those it is well known that there is more likely to be mental disturbance, showing itself by loss of emotional and other inhibitory control. It is, however, quite possible that the lesion itself is irritative, but not always of itself sufficient to produce the phenomena of Chorea, unless the inhibitory powers are weak as in the child, or weakened as by the presence of pregnancy.

Theories as to the causation of Chorea have naturally not been wanting, but we are gradully approaching to an agreement on this subject. We shall discuss here three views, namely:-

1. That it is due to an infection, and that its chief symptoms are the result of a toxaemia.
2. That it is a "functional" disease.
3. That it is due to multiple minute emboli.

1. Infective Theory:

The view that Chorea is one of the ordinary infective/

fective diseases, has long been considered and has been rapidly gaining ground during the last few years.* Sydenham was unconsciously very near the truth, when he wrote that "now this affection arises from some humour falling on the nerves; and such irritation causes the spasms." One of the most striking facts in the disease is that it is one of great variations in severity. We have very mild cases where there is merely the inconvenience caused by the Choreic movements, and these may be slight, and where there is no interference with the child's general health up to those very severe and acute cases of Chorea Insaniens, which may prove fatal in a few days. It is the occurrence of those very mild cases which may make one hesitate to put them in the same category as those severe ones, which correpond in every detail with an acute infection. Another of the chief features of Chorea is that it is mainly a disease of children, and that it largely predominates in the female sex, and that in adults it is apt to attack women during pregnancy. Besides this, it is especially prone to attack children with some degree of mental instability, whether congenital as in cases with a neurotic heredity, or acquired, as in overworked school children/

* Works of Sydenham - Latham Schedula Monitoria, para. 19.

children. But all those are facts which are not in any great degree against the infective theory of the disease; the relatively greater invulnerability of a certain class or age is one of those problems of immunity which will be solved some day. The study of immunity, which has made such rapid strides of late, will have a great deal to do with the clearing up of our views on the subject of Chorea, as of many other diseases. The influence of fright in the causation of Chorea, though to my mind exaggerated, is undoubted and may be difficult to reconcile with the infective theory. In no other infective disease is fright usually considered one of the important factors, but, as Gowers has shown, fright is an emotion that has a profound influence on the motor system through all classes of the animal kingdom, and possibly the shock to the motor cells may so alter their nutrition that they may be less resistant to the infection. The presence of Endocarditis in such a large proportion of cases is a most significant fact, as endocarditis is in most cases an infective condition. It is found in many diseases, e.g., Rheumatism, Gonorrhoea, Typhoid, Scarlet Fever, Septicaemia, etc., all of which are organismal diseases. There, therefore, seems to be every/

be every reason to include endocarditis in Chorea under the same heading, for it differs neither in its Morbid Anatomy, nor in its course from that found in the afore-mentioned diseases.

Similarly, Pneumonia, Pleurisy, and Nephritis are sometimes found in Chorea and they also point to a morbid blood state.

In the sections of Etiology and Morbid Anatomy, something was said of the association of Rheumatism with Chorea, and we must now say a few words more on this subject.

Now what is this association, for it is undeniable that the diseases are related? The results of recent research on the bacteriology of Acute Rheumatism have been to establish the claims of a specific diplococcus to be at least a cause of the disease, and several observers have recently found an identical organism in the brain and pia mater and on the heart valves of cases of Chorea. Inoculation experiments also, to some extent, are in favour of the view that the organism is the same in both diseases. Are we then to believe that the diseases are the same, that Chorea is merely one form of "cerebral" Rheumatism, perhaps specially modified/

fied by its occurring in a child? Sir Thomas Barlow* long ago pointed out that many children suffering from Rheumatic Fever were very nervous and excitable, while others showed very slight traces of Chorea. Undoubtedly, Rheumatism differs in a child from the same disease in the adult, but the evidence is as yet hardly full enough, nor has it been sufficiently confirmed for us to come to this conclusion. It may be that Chorea can be produced by other causes; it may be that it is a functional disease occurring in patients whose cerebral stability is from one cause or another impaired, and that Rheumatism is only one of many causes leading to damage of the cerebral cells.

Another view is that Rheumatism is not due to any special organism, but to a combination of several and that a preponderance of one of those may give rise to arthritis, and of another to Chorea. This is quite a plausible theory, but it must be noted that it is a theory only and has no established basis of fact.

Though a dogmatic opinion is not possible at present, I believe that future research will show more clearly the identity of the two diseases.

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* Quoted in Lancet, May 4, 1901, p. 1264.

2. "Functional" Theory:

The chief supporter of this view was the late Dr Octavius Sturges. Some of the facts in the life history of Chorea are certainly a little difficult to reconcile with the idea of an organic disease, but on the other hand, many are impossible to reconcile with any other hypothesis.

The importance of fright as a cause of Chorea is insisted on by those who are in favour of the functional nature of the disease, but I believe that its influence is indirect, as was mentioned in the preceding section.

Similarly, the special frequency of the disease in females and especially in girls about the time of puberty and during pregnancy, and its special frequency in patients with more or less unstable nervous system are adduced in support of this view. But for a disease to have a special tendency to affect one sex and to be commonest at a certain age, is quite usual and does not negative its toxic nature. It is quite likely that an infection with the Micrococcus Rheumaticus, which would produce an arthritis or endocarditis at one time, or which might never produce any pathological condition at all, may give rise to Chorea where the cerebral/

cerebral cells are rendered vulnerable by their condition at those epochs of puberty or pregnancy.

Sturges pointed out that an excitable and restless child may make movements like those of Chorea and believed that Chorea was merely "an extravagant exaltation of that continual unrest which is the natural characteristic of childhood,"* but this does not prove the disease to be merely functional. It is only natural that an irritative lesion of the cerebral cells should result in movements, etc., more or less resembling those that are normal.

The tendency to recurrence has been cited in favour of the functional nature of the disease, but surely Acute Rheumatism is equally given to recur and no one considers that a functional disease.

Chorea is a disease with a more or less definite course, a beginning, a middle, and an end; its average duration is ten weeks and if it were merely a functional disturbance due to fright, etc., occurring at a time when the motor centres were in a stage where there "functional development was complete, but their exercise not yet stable" (Gowers), onw would not expect that a period of ten weeks would/

* Sturges, loc. cit., p. 15.

would so alter this as to lead to entire cessation of the movements. No, that epoch must extend over a much longer period, and the cessation of the choreic movements is due not to this stage having been completed and rendered stable, but to the destruction of the active cause by those processes, antitoxic and phagocytic, etc., whereby the body is able to destroy pathogenic bacteria plus the time required for repair. Hysteria is a disease of whose nature we know little; it is generally classes as a functional disorder and its claim to be so is much more valid than that of Chorea, though possibly, future research may find another classification for it. It occasionally is found along with Chorea but that is no reason why we should place the latter under the same heading.

Coming next to the occurrence of Endocarditis, no amount of explaining on a functional basis can explain that away, and so also the association of Chorea with Rheumatism is equally inconvenient from the functional point of view. Lastly, supposing we do adopt the "functional" view, are we really so very much the wiser? Dr Dturges was aware of this objection and could say little to lessen it. In concluding a somewhat spirited attack on the then/

then current views on the pathology of Chorea, he says: "Impaired nutrition of nerve centres; disruption of the normal relations between such centres; excessive, defective, or misdirected nervous discharge by cells of some particular part of the brain cortex, these and similar expressions, commit us to nothing, and in so far as they have a precise meaning, may find ample justification some day."* If he had included "functional disturbance" in this list, it would have been still more complete.

3. Embolic Theory:

We shall only say a few words on this subject as, though the theory was once strongly supported, it has now become practically obsolete. It was first originated by Dr Kirkes, who suggested that the Choreic movements were the result of showers of minute emboli, more especially in the region of the corpus striatum. The objections to this theory are overwhelming.

In the first place, it has received very little support from morbid anatomy. Emboli do occasionally/

* On Chorea, Sturges, 2nd Edit., p. 134.

occasionally occur in Chorea and minute vessels in the brain have been found occluded by them, or by thrombi in several cases examined post mortem. But this is not one of the commoner post mortem findings, and considering the frequency of endocarditis in Chorea, it is astonishing how seldom embolism is found. Besides, Endocarditis is not invariably present, so that a separate pathology would be required to explain those cases.

Experiments by Money gave only slight support to the embolic theory. He injected minute particles into the carotids of dogs and found that Choreiform movements were sometimes produced, but only when the particles reached the upper part of the cord. The other phenomena of Chorea were not seen.

Chorea does not commonly arise in the course of Rheumatic Endocarditis in the adult, nor in ulcerative endocarditis where embolism is much more frequent. In view of those facts, it is obvious that the theory is untenable.

TREATMENT:

We shall consider firstly the question of prophylaxis. This, except in the prevention of second attacks, is a measure that we rarely have opportunities of taking. Chorea being a disease apt to occur in children with a neurotic or rheumatic inheritance, and being in some instances brought on by nervous shock or strain, it is obvious that we should endeavour to protect such children from any cause tending to increase their mental instability. We must see that they are well clad, well fed, not overworked at school, not over-excited at games, and so on. Of course, in many case these are counsels of perfection impossible of attainment. In rheumatic children, timely treatment of any rheumatic manifestations by means of Salicylates, etc., may be of value in preventing the onset of Chorea.

With regard to second attacks, some writers have attempted to demonstrate a tendency to vernal recurrences, and administer arsenic in tonic doses at that time, but of course, the evidence of its value is impossible to obtain.

Taking next the <u>actual</u> <u>treatment</u> of a case, it may/

may be divided into two sections:-

1. General treatment,
2. Drug treatment.

1. General Treatment:

The factor of pre-eminent importance is rest. All cases of Chorea, however, mild, should be treated by rest in bed throughout the greater part of the illness. Under no other treatment is the disease so rapidly cured, and with rest alone, we can cure the greater number of cases without resorting to drugs at all.

When we consider, too, the enormous proportion of cases that develop endocarditis and that this complication has no marked relation to the severity of the attack and that it may be present without recognisable physical signs, it is evident that here is a very evident reason for careful treatment. The statistics of Osler already cited with regard to the condition of the heart some years after an attack, are very striking and ought to teach us that, though Chorea may be rarely fatal, it is often the starting point of life-long cardiac disablement. No one thinks of letting a mild case of Acute Rheumatism/

atism go about, yet how often is a child with Chorea treated at the out-patient department of a hospital! All are agreed that heart complications are rendered less likely by strict rest.

Apart from the question of Endocarditis, we have seen that voluntary movements tend to aggravate the choreic, so here is a further justification for our treatment. The prejudices of the laity against the "weakening effects" of prolonged rest in bed are hard to contend with, more especially as the patient may feel very well, but a strong effort ought to be made to get it carried out with thoroughness.

Besides physical rest, the child must have mental rest and this is best secured by isolation. To a nervous sensitive child, the consciousness of its movements being observed and probably caricatured by its unsympathetic younger brothers and sisters is very distressing and is apt to increase the Chorea. The child should then be kept in bed; her surroundings ought to be pleasant, but she must not be excited. A trained nurse is a great help, as the child's mother is not, as a rule, the most suitable attendant. In cases of greater severity, care must be taken to prevent the child injuring herself/

herself; thus, the prominent parts of the body may be covered with cotton wool, the bed may be so padded as to prevent injury, and in very violent cases, the mattress may be laid on the floor. A water bed is often useful.

Food: Food ought to be nourishing and simple. An exclusively milk diet given in large quantities has been recommended by some, but is by no means necessary or called for.

In severe cases, great difficulty may be experienced in feeding, as the child's violent struggles render it almost impossible to get food administered, and besides which, choreic movements of the mouth and tongue may interfere with the act of deglutition. In such cases, food must be, of course, fluid and may be administered either from one of the "tube" variety of baby's bottle, or from an ordinary feeding cup with a piece of rubber tubing attached to the mouth piece. Occasionally recourse has to be had to rectal alimentation.

Massage: Has been tried in some cases, but is of no very special value. It is most likely to be of service in chronic cases.

Electricity/

Electricity: In any of its forms is also of little use.

Baths: Prolonged warm baths are strongly recommended by Jacobi* in mild cases, and in more severe ones, where this is impracticable, warm sponging is very useful and soothing. In cases of long standing, where only slight Chorea remains, cold baths or cold douching to the spine are exceedingly useful.

Change of Air: After an attack, a change to the country or seaside, when obtainable, should be recommended, but one must caution the child's parents against allowing any undue excitement or exertion. Ashby** lays special stress on this and says that if a chronic case is sent off before the Chorea has entirely ceased, it is very usually aggravated.

2. Drug Treatment:

On none of the sub-divisions of this subject of Chorea are opinions more widely divergent than on this. From the optimistic view of one writer*** who/

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* Therapeutics of Infancy, Jacobi, 3rd Edit., 1903, p. 298.

** Diseases of Children, Ashby & Wright, 4th Edit., p. 526.

*** "Rough Notes on Remedies", Murray, 4td Ed., p. 18.

who says that in nearly twenty years he has seen only one case which resisted treatment by arsenic, down to that of Sturges and many others, who hold that no drug is of any value as a curative agent - we have all possible gradations. Then, we have the supporters of other drugs than arsenic, of Ergot, of Antipyrine, of Chloral, of Salicylates, etc., all equally anxious to show that their favourite remedy is never-failing. Truly, a complete list of the drugs that have been vaunted as specifics, is a formidable affair, and one wonders how Chorea can still be a disease with an average duration of ten weeks, but no doubt the old saying about too many remedies for one disease is true here as elsewhere.

The main reason for all this diversity of opinion is that it is almost impossible to definitely prove that the recovery of a case is due to the remedy employed. The disease is one that tends to get well spontaneously, and it has not a very settled duration; thus, one case may get well in a few weeks, whilst another, at first apparently of similar severity, takes months. The most satisfactory way to decide on the value of any special drug would be to employ it in a large number of cases and see if the average duration is shortened./

shortened. Although this has been done, it has not been carried out on a sufficiently large scale to make the results trustworthy.

Another fairly good test of the therapeutic activity of a drug, is to try it in a series of the more chronic cases; if its administration is repeatedly followed by improvement, we are justified in concluding that in all probability it is due to our remedy. Another source of difficulty in determining the efficacy of drugs, is that there is great difference in different cases in their response to certain drugs. Thus we may find that arsenic is of use in one case, and inert in another; so also antipyrine useful in one case, and inert in a second, and so on. We may broadly divide the various drugs used in Chorea into three groups.

1. Hypnotics, or motor depressants, e.g., Chloral, Chloroform.
2. Anti-rheumatics, e.g., the Salicylate group.
3. Empirical (or "alteratives", if it is preferred), e.g., Arsenic.

1. Hypnotics:

There is no question as to the value of hypnotics at certain times and in certain cases; there is/

is considerable room for doubt as to the advisability of using them as a routine treatment.

In very severe cases where the movements are incessant and in cases of Maniacal Chorea, hypnotics are urgently required. Jacobi* advises keeping very bad cases asleep by means of hypnotics for eighteen out of the twenty-four hours. As we have seen, the choreic movements cease during sleep, but where their severity is such as to prevent sleep, it is often very useful to give a dose of Chloral at bed time.

Of the various hypnotics most used in Chorea, Chloral, Opium, and Trional are the best.

Of these, I believe Opium to be the best in very severe cases. Small doses are said to be excitant, but large doses are very effectual, and are no more dangerous than similar doses of Chloral or other narcotics. In my opinion, the dangers of opiates given to children are exaggerated. It may be given as Morphine by the mouth, or occasionally hypodermically.

Chloral is strongly recommended by some and is of great use in many cases, but its cardiac depressant action renders the use of such large doses as are/

* Jacobi, loc. cit., p. 297.

are sometimes required to produce sleep, not free from risk in a disease where cardiac involvement is so usual. It is, however, probably the best remedy for general use in milder cases, where moderate doses are sufficient. Some writers have advised its routine administration as a more or less specific remedy, but I have never found it so. Gairdner has recorded an interesting case, however, where a child accidentally swallowed a drachm of Chloral Hydrate and suffered from toxic symptoms, but was at the same time cured of her Chorea.

Trional has also been much advocated of late years, but in my own experience, has no advantages over some of the older narcotics.

Bromides are generally regarded as being of less use in Chorea than in some other diseases where excessive motor discharges are the chief pathological condition, e.g., Epilepsy. This is a point of some theoretical interest. I have, in some cases, used Bromides along with Arsenic with good results.

Chloroform is sometimes an indispensable aid in those very severe cases with incessant movements, and in which the child is rapidly losing ground from/

from pure exhaustion; while under its influence, one can administer such remedies as opium or chloral by the rectum. Often the child will fall into a natural sleep from some hours after the administration of Chloroform alone.

Hyoscine has been very little used in Chorea, though one would expect that it would be useful. It is, however, a powerful drug and not quite free from danger. Starr* recommended it in severe cases.

Conium has now in great measure fallen into disuse and having regard to its chief pharmacological action of depressing the activity of the motor nerves and its entire absence of action on the brain, it can easily be understood that there is very little rational indication for its use, and in fact, that it is attacking the disease from the wrong end.

Cannabis Indica, Monobromated Camphor, etc. are all occasionally employed, but are of little importance.

2. Anti-Rheumatics:

In view of the close relationship between Rheumatism and Chorea, one would naturally expect that/

* American Textbook of Diseases of Children, Starr, 2nd Edit., p.

that anti-rheumatic treatment would be as effectual in the latter as in the former, but the experience of most clinicians leads them to a contrary conclusion. Of course, it must be remembered that therapeutists are not agreed as to the Salicylate group having any decided action over and above being pain removers; it has not been decided whether they should be regarded as having any power to prevent say, - endocarditis or hyperpyrexia, etc. Most probably they have some action in this direction, but it is less marked than the very obvious action in relieving the pain due to the inflamed joints of Rheumatic Fever. Wild* believes that Salicylic Acid compounds have a specific action on the rheumatic toxins, as similar good effects are not obtained by the use of such bodies as meta-, and para-hydroxyl benzoic acids, which are isomeric with Salicylic Acid. If one then gives Sodium Salicylate in ordinary doses to a choreic child, one usually finds but little improvement. Lately Dr Lees** has been endeavouring to show that provided "adequate" doses are given, the action of the Salicylates is just as beneficial as in Acute Rheumatism. He bases his theory on the analogy of giving large doses of Potassium Iodide in Syphilis of the central/

* Brit. Med. Journal, Jan. 3, 1903.

** Brit. Med. Journal, Aug. 29, 1903.

tral nervous system, and this analogy is to some extent valid.

In a child of six to ten years, he advises that Sod. Salicylate be given in gr. X. doses two hourly during the day and three hourly at night, that is, ten doses in the twenty-four hours. After a few days, the dose is increased to gr. 15 and in a few more to gr. 20. As such large doses are not free from danger, he advises the combining with each dose of the Salicylate of a double quantity of Sod. Bicarbonate, believing that the alkali is able to ward off symptoms of Salicylate poisoning and especially that form which he calls "Air Hunger". This is occasionally seen where large doses of the Salicylates are given alone, and very much resembles diabetic air hunger and is in all probability due to some destructive action on the red blood corpuscles. The treatment with alkalis probably also lessens the chances of endocarditis. Dr Lees states that the ordinary disagreeable symptoms occasioned by large doses of the Salicylates, e.g., noises in the ears, headache, giddiness, etc., are rare in children and that the only difficulty in his treatment is that it may cause vomiting. This, he says, may be obviated by dropping out a few doses and then/

then beginning again with a somewhat smaller dose.

Well, treatment of this nature borders on what is usually termed "heroic", and has not, I believe, found universal favour. In my own experience, I have not noticed any immunity of children to Salicylism, and I can hardly imagine that a child of from 6 - 10 years of age who is consuming daily for a period of at least a week (as Dr Lees advises) a total of from 100-200 grains of Sod. Salicylate should escape from it, and as any one who has experienced it can tell, the symptoms are exceedingly unpleasant and may be dangerous.

Dr Lees is of opinion that the salicylates have little or no depressing action on the heart and that all cases of heart failure during their administration are due to rheumatic dilatation of the left ventricle. This, I venture to believe, is by no means the opinion of the great majority of practitioners. The Salicylate group have been proved by definite physiological experiments to have such a depressant action when given in large doses. Wood* states that "the final fall of the arterial pressure (in toxic doses) is in large part, if not altogether, due to a direct action upon the heart itself." He also mentions that small doses cause some/

* Therapeutics, 11th Edit., 1902, Wood, p. 578.

some increase in blood pressure, probably by stimulating vasomotor centres. It will thus be seen that the use of such large doses in a disease where dilatation of the heart is prone to occur is by no means free from danger.

It is probable that smaller doses, say, up to 3i daily will be quite sufficient if the case is going to respond to anti-rheumatic treatment at all. No doubt, the addition of alkali may be in one way an advantage, but the counter disadvantage of repeatedly distending the stomach with CO_2 must be borne in mind, and thus it may be better, as Ewart recommends, to administer Potassium Citrate in place of the Sodium Bicarbonate.

Williamson* has lately testified to the good results obtained in Chorea by treatment with Aspirin, one of the newer members of the Salicylic Acid group, which is said to have none of the disadvantages of the ordinary acid.

Dr Williamson administered it in fairly large doses to thirty-five consecutive cases. These were all treated as out-patients and though kept from school, had not the advantages of rest in bed. He came to the conclusion that though not a specific, aspirin had a very favourable influence on the course of/

* Lancet, Aug. 22, 1903.

of the disease. In no case was there absence of improvement and this, although amongst the number, were several old and chronic cases which had previously been treated with other remedies. Absolute proof is, of course, not to be had, but taken in conjunction with Dr Lees advocacy of Salicylate treatment, one would be inclined to believe that Aspirin has a distinct value and is in many respects preferable to correspondingly large doses of Sodium Salicylate.

3. "Alteratives":

We now come to a very large number of drugs which have been used and recommended in the treatment of Chorea. These mainly belong to that vague class known as "alteratives", and their use in Chorea is at present in great measure purely empirical.

The most important of this group is arsenic.

Arsenic: This is probably the most used of all the many drugs used in this disease and the difference of opinion as to its efficacy is most extraordinary. The physiological action of Arsenic is/

is little understood; there is not doubt that it has in some cases a powerful action on metabolism, but what exactly this is and how it is produced, is not known. In toxic doses it has a distinct action on the nervous system, the terminal coma and convulsions being due to a direct action on the cerebrum.

In all probability the diversity of opinion among physicians as to its use in Chorea is due to lack of agreement as to the dosage. Dr Murray of Newcastle*, the most ardent supporter of the Arsenic treatment, insists on at least fifteen minims of Liquor Arsenicalis being given to a child thrice daily and that this dose be kept up for a week. If there is no improvement at the end of that time, the Arsenic must be stopped, as toxic action would be produced. Dr Murray goes so far as to say that he has only seen one case in nearly twenty years, which has resisted this treatment, and without going so far as this, I myself believe that if given according to this plan, a very large majority of cases will improve and in some the rapidity of improvement is very striking. The following is an illustrative case:-

Emily C: age 10, was admitted October 29th, 1902/

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* Murray, loc. cit., p.18.

1902 with moderately severe Chorea. This was a second attack and had commenced rather more than a month previous to admission. She also complained of being easily tired and was dyspnoeic on exertion. Her heart was slightly enlarged and on auscultation there was a distinct short mitral systolic bruit and a slight "blow" after the second sound. There was no presystolic. The systolic murmur persisted during patient's stay in hospital, but the diastolic faded.

The patient was put on Liq.Arsenical: in small doses which were increased at the end of six days to gr. 12 . The improvement was most rapid and by November 10th, she was able to write her name quite distinctly.

The Arsenic must, of course, be given after, or during a meal and well diluted. I have been in the habit of beginning with gr. ii doses and doubling the dose every second day; in a few cases, I have begun with gr. 15 straight away, but this is perhaps rather more likely to cause vomiting or diarrhoea.

Toxic symptoms, such as vomiting and diarrhoea, watering of the eyes, an erythematous rash, etc., of course call for a reduction in the dose, or an entire cessation. I have seen no instance of neuritis/

neuritis following this treatment, excepting slight Herpes in two cases, which may have been due to something of the nature of a neuritis. The symptoms in the following interesting case I can only explain on the theory of an exceptional intolerance to arsenic.

F.C., male, age 11. Admitted in his second attack of Chorea on May 14th, 1902. He had had Rheumatic Fever two months previously. On admission had a severe grade of Chorea with a good deal of paresis of the left arm. No heart lesion.

On May 16th, he was put on Liq. Arsenical. gr. iii. T.I.D. On May 19th began to complain of severe abdominal pain and vomited frequently. On May 21st, though the Arsenic had been stopped for two days, the vomiting continued and there was now also severe diarrhoea. Nothing abnormal was to be felt in the abdomen. On now examining the heart, a rough mitral systolic murmur was to be heard. This remained as long as patient was under observation. The diarrhoea continued for four days and then ceased. The chorea, during the attack of vomiting and diarrhoea, improved. Patient was discharged quite well on July 3rd.

The hypodermic use of Arsenic in bad cases where/

where one desires to get the patient quickly under the influence of the drug, has been recommended and may be useful in adults, but in children, for obvious reasons, is less desirable. In severe cases, too, as of Chorea Insaniens, Arsenic is not so likely to be of service as in the more ordinary ones, though Pope* states that he has occasionally had remarkably good results.

Antipyrine: The good results sometimes seen after the administration of Antipyrine in Chorea were first pointed out by Comby of Paris. The precise physiological action that makes it of service is not clear. Its antipyretic and analgesic action in other diseases show it to possess a distinct action on some part of the nervous system. It must be given in large doses to have any effect. In a child of from six to ten years of age, one would give up to 10 grains four hourly, or even occasionally gr. XV. I have never seen any bad results from such doses other than in one case a very extensive "measly" rash, but signs of cardiac weakness ought to be looked for. If albuminuria appear, the drug must be stopped. The temperature often remains slightly subnormal during the administration.

Wood/

* Brit. Med. Journal, Oct. 18th, 1902, p. 1229.

Wood* says that Antipyrine is eliminated more quickly in children than in adults and this probably explains their freedom from bad effects.

I believe that next to Arsenic, Antipyrine is the most valuable of all the many drugs used in Chorea.

The following case shows well the good results of treatment with Antipyrine.

E.R., Female, aged 10½. Admitted in her second attack on December 16th, 1902. There was no rheumatic history. The present attack began four months ago. On admission she had a moderate degree of Chorea. There was some cardiac dilatation but no bruit. She was given ordinary diet, and Antipyrine gr. V. four hourly, increased after two days to gr. X. four hourly. This dose was continued for four days, at the end of which time there was no trace of Chorea. She was discharged perfectly well on January 8th.

Exalgin is much used in America, but I have no personal experience of it.

Ergot: Dr Eustace Smith** has recently published a paper strongly recommending Ergot in very large/

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* Wood, loc. cit., p. 590.

** Brit. Med. Journal, Vol. II., 1903, p. 133.

large doses. He advises that drachm doses of the Liquid Extract of Ergot be given to children of seven or eight, and that this should be given every three or four hours and continued for some weeks if necessary. In some cases Dr Smith has given considerably larger doses, but the above he regards as generally sufficient. The pulse usually falls from fifteen to twenty beats per minute in rate, but he has never seen any bad results. Latterly, he has combined the Ergot with small doses of Strychnine and considers this an improvement. In my own experience of a few cases treated on these lines, I have failed to see that any decided benefit was produced, nor can I quite agree as to the absence of untoward symptoms. In several cases the pulse, besides being much slowed, became exceedingly feeble and to some extent irregular.

Dr Smith does not offer any explanation of the pharmacological action of Ergot in such cases other than to say that it acts as a sedative, or possibly as a vasoconstrictor on the vessels of the cord. Ergot is generally considered to have very little action on the nervous system and as far as my own experience goes, this is fully borne out by the results of its use in Chorea.

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A large number of drugs remain, but it is unnecessary to do more than mention a few of them.

Cimicifuga is used to some extent in America and Wood states that its value is unquestionable.

West* recommended hot air baths and a dose of Antimonial Wine to be followed up later by large doses of Sulphate of Zinc.

Physostigmine, Belladonna, and even Curara have been used, but their practical value is nil.

In chronic cases, Strychnine is sometimes of service. Ine one extremely chronic case with marked mental hebetude which had been dragging on for months, I went through the whole gamut of remedies, Arsenic, Antipyrine, Cannabis Indica, Opium, etc., without the slightest improvement. Thyroid Extract made the patient rather more lively, but had no effect on the Chorea and it was ultimately left to an accidental infection with Scarlet Fever to modify for a time, at least, the movements.

Serum Treatment:

The serum treatment of Acute Rheumatism is as yet only in the experimental stage, but the results especially in chronic cases, have been encouraging. Menzer/

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* Disorders of Nervous System in Childhood, Charles West.

Menzer,* working with a serum produced by immunising animals with streptococci from the tonsils of rheumatic patients, found that the duration of an acute attack was shortened, though the serum did not relieve the joint pains as Salicylates do. Endocarditis also appeared to be less frequent.

This serum has so far been only used in one or two cases of Chorea and no sufficient data are as yet available.

Lumbar Puncture:

Jemma** records two severe cases of Chorea treated by this method. There was immediate improvement, but relapse occurred, which was again relieved by a second puncture. Possibly this method of treatment may be useful along with treatment by arsenic or other drugs.

To sum up the treatment, we should say that the great essential is rest, mental and bodily. Isolation from over-indulgent parents and from heartless young brothers and sisters is often the first step towards recovery.

Next to that in importance is a simple nourishing diet/

* B.M.J. Epitome, Sept. 27, 1902, p. 47.

** Do. Jan. 25, 1902.

ing diet. Of drugs, Arsenic and Antipyrine are the most useful and their beneficial action in some cases is certainly very striking. In very severe cases, sleep must be procured by means of hypnotics. In these cases the patient's strength must be kept up by liberal feeding and this is often a matter of extreme difficulty.

In chronic cases, gentle exercise, massage, and mild gymnastic exercises are useful. So also are cold bathing or douching. It is unnecessary to enter into the question of treatment of such complications as pericarditis, etc; these must be treated on ordinary lines.

It is interesting to note that nearly all the drugs most recommended in Chorea have to be given in doses which their respective advocates mildly call "adequate", but which might more fittingly be described as overwhelming. Indeed, as Sturges was disposed to think in regard to arsenic, it was only a question of making the patient feel ill enough for one to cause a cessation of the movements. This, however, seems hardly to be the whole truth, for if it were,one would expect the Chorea to recommence very soon after ceasing those large doses, but this is/

is by no means the rule; still, this suggestion has, I think, some foundation in fact.

In conclusion, I have to acknowledge my indebtedness to Drs Ashby and Hutton of the Manchester Children's Hospital for their very cordial permission for me to make use of their cases.